Paciente masculino de 26 años, etilista y consumidor de cocaína que presenta bradicardia y paraparesia de los cuatro miembros – 2010

Dr. Edgardo Nuñez

Colegas del Foro:

A continuación comentaré una Historia Clínica, de un paciente vivido en el día de ayer, y agradecería alguna opinión al respecto:

Se trata de un paciente del sexo masculino, de 26 años de edad, que como antecedentes a destacar presenta: fumador intenso, etilista, drogadicción con alto consumo de marihuana y fundamentalmente cocaína. El día 15/2/02 presenta picaduras de abeja a nivel de cabeza presentando reacción alérgica, que es tratada con fármacos antihistamínicos. Dos días después comienza con astenia (durante estos días y según relato de familiares, existe gran consumo de cocaína).

El 18/2/02 ingresa al Sanatorio por paraparesia de los 4 miembros a predominio proximal.

Visto por neurólogo plantea probable poliradiculoneuropatía, ingresándolo y realizándole una punción lumbar que fue normal.

Le constata una bradicardia por lo que consulta a cardiólogo, quien realiza ECG, el cual muestra ritmo nodal de 38 p.m., solicitando ingreso a la Unidad Coronaria para monitoreo y eventual implante de marcapado transitorio. En ese instante tomamos contacto con el paciente,el cual estaba estable, destacándose del examan la paraparesia a predominio proximal, con reflejos conservados. El ECG mostraba ritmo nodal de 40 pm, que con 1 mg de atropina i/v pasa a un ritmo sinusal de 77 pm, quedando posteriormente con una bradicardia sinusal de 40 pm, alternando con ritmo nodal.

CK total de 110 con MB de 12, test de troponina T negativo. Ecocardiograma doppler normal.

Es conocido la asociación de eventos coronarios, arritmias rápidas, supra o ventriculares con el alto consumo de cocaína, pero no el de ritmos lentos, con escaso aporte en la literatura consultada.

Agradezco vuestras valiosas opiniones.

Gracias

Dr. Edgardo Núñez

Punta del Este-Uruguay

OPINIONES DE COLEGAS

Prezado colega Dr. Edgardo Nuñez: tenta lhe responder Andrés Ricardo Pérez Riera de SP Brasil.

O jovem de 26 a drogadicto por cannabioides e cocaina apresentado tivera uma reação alérgica por picadura de avelha e le fora administrado um antihistamínico. Seria para nos interesante saber qual droga foi administrada.

Seria um antihistamínico de primeira geracao? (etanolaminas, etilendiaminas ou alquilaminas)

Seria um antihistamínico de segunda geracao? (alquilaminas, piperazinas ou piperidinas) uma vez que os antihistamínicos podem ocasionar reacoes importantes no coração e no SN. Por Ex o não sedativo antagonista H2 terfenadina (piperidina) e o anti-histamínico antagonista H1 de ação longa astemizol, (piperidina) recentemente suspensos pela Vigilância Sanitária em nosso País pelo seu potencial de originar grave arritmia (TVP) se administrado acima das doses recomendadas ou quando em interação com outras drogas (neste caso cocaina). A proibição dos medicamentos à base de Astemizol foi determinada em função do risco de arritmia cardíaca que representa para os pacientes quando tomados acima da dose permitida ou associados a outras drogas como claritromicina, mibefradil, troleandomicina, eritromicina, cetoconazol, itraconazol e quinonas.

Os Astemizol, isolado ou em associação, está presente em anti-alérgicos usados no tratamento de rinites, conjutivites, urticária e prurido. A Agência sanitária em nosso pais

(Brasil) já havia divulgado alerta sanitário avisando médicos e farmacêuticos sobre o risco de associações e do mal uso de produtos à base de astemizol.

Em referencia ao quadro neurológico lembre que os antagonistas H1 de primeira geração podem tanto ocasionar um quadro excitatorio quanto inibitório com sonolencia, reacoes mais lentas e isto associado a coca pode se potencializar (interacao). Por outra parte, alguns antihistamínicos possuem efeito anestésico local que bem poderia justificar o quadro transitorio de paraparesia dos 4 membros.

Se se trata de antagonista H1 de segunda geracao estes não tem efeitos sobre os receptores muscarínicos. Finalmente a maconha (marihuana em espanhol) pode ocasionar trastornos da coordenacao. Os sinais da supresao da cocaina inclui bradicardia. Pode o paciente ter tido uma intensa reacao vagal compensatória. O parasimpático a cargo do vago ou X par craniano no coração tem inervação direta predominante em nódulo sinusal, átrios, sulcos A-V e sistema de condução alto sendo sua ação nos ventrículos muito ténue.

Seu mediador a acetilcolina (Ach) em concentração de 10-5 não ocasiona nenhum efeito no endocárdio e efeitos pronunciados no PAT do epicardio. Em baixas concentrações (10-6 a 10-7) produz prolongamento do PTA e em concentrações maiores encurtamento. O efeito é bloqueado pela atropina. A ação de acetilcolina na duração do PAT obedece a inibição do canal ICa++ (Fischmeister, R.; Hartzell, H. C.: Mechanism of action of acetylcholyne on calcium current in single cells form frog ventricle. J. Physiol. (London) 376:183-202, 1986) ou por ativação do canal Ik-ACh ou Acetylcholine-activated inward rectifyin current Canal de K + ativado pelo receptor muscarínico M2 importantes no nódulo SA, nó AV e átrios.

Nos dois primeiros, podem produzir hiperpolarização e pode ocorrer ritmos de suplencia juncional como acontecera no seu caso e nós átrios encurtar o PTA. Nos ventrículos encontra-se apenas nas células de Purkinje estando ausente nas células banais ventriculares.

Quanto a cocaina como voce bem comentara, pode induzir a infarto agudo ou predispor a ateroesclerose coronariana fato que não ocorrera no caso apresentado, porem, vale a pena comentar quais são os mecanismos pelo qual cocaina leva a infarto e ICo:

1) apenas 10' após a ingestão inalatória ou venosa produz-se intensa liberação de adrenalina, esplenocontração, aumento da taxa de glóbulos vermelhos em 6% conseqüente aumento da viscosidade sangüínea o que predispõe a trombose.

2) em apenas 30' verifica-se aumento de até 40% do fator de Vom Willebrand conhecida proteína que facilita a aderência plaquetária.

3) aumento na frequencia de trombose coronariana sem hemorragia da placa.

4) espessamento fibrointimal do endotélio coronariano (Pamplona, D.; Gutierrez, P. S.; Mansur, A. J. et. al..: Fatal acute myocardial infarction in a young cocaine addicts. Arq. Bras. Cardiol. 55:125-7,1990).

5) vasoespasmo (pela via inalatória) mais intenso nas áreas com placa ateroesclerótica (Flores, E. D.; Lange, R. A.; Cigarroa, R. G. et. al.: Effect of cocaine on coronary artery dimensions in atheroesclerotic coronary artery disease: enhanced vasoconstriction at sites of significant stenoses. J. Am. Coll. Cardiol. 16:74-9, 1990).

6) maior incidência de infarto agudo e morte tanto em usuários em forma aguda quanto crônica. O risco de uma pessoa sofrer infarto agudo após cheirar cocaína aumenta 24vezes. Esta maior tendência a infarto obedece à seguinte seqüência: taquicardia, maior requerimento de oxigênio por parte das células cardíacas, aumento da viscosidade sangüínea, liberação do fator de Von Willbrand maior adesividade plaquetária, espasmo arteriolar, lesão miointimal, trombose... infarto.

7) mais de dois terços dos usuários de cocaína que desenvolvem infarto agudo apresentam concomitantemente dois fatores de risco para doença arterial coronariana (Hollander, J. E.; Shih, R. D.; Hoffman, R. S.et. al.: Predictors of coronary artery disease in patients with cocaine-associated myocardial infarction. Cocaine-Associated Myocardial Infarction (CAMI) Study Group. Am. J. Med. 102:158-63, 1997).

Andrés R. Pérez Riera

En la bibliografía a través de Medline se puede apreciar reportes de bradicardia por intoxicación con cocaína, síncope en jóvenes en los cuales la causa fue la misma. Disminución del output simpático central y posibles acciones colinérgicas directas podrían ser la causa.

Por los antecedentes relatados de este caso en particular debe ser un muchacho no muy bien nutrido. Recuerdo un caso de un joven con bradicardia muy importante, sin respuesta cronotrópica adecuada, con desnutrición proteica. Dicha bradicardia excesiva desapareció luego de un buen plan alimentario.

Dr Jose Luis Serra.

Cordoba

Disculpen mi intromisión cuando la pregunta se refiere puntualmente a la arritmia, pero al leer el cuadro lo primero que pensé fue en la posibilidad, aunque remota por lo infrecuente, de que el paciente haya desarrollado un hematoma epidural, el cual podría explicar el cuadro neurológico. Si fuera el caso, el nivel medular como mínimo debería ser torácico alto, involucrando el simpático cardiotorácico. Las lesiones medulares a nivel T1 a T5 cursan con bradicardia y marcada inestabilidad hemodinámica.

Adjunto dos citas que mencionan hematomas epidurales espontáneos.

Seria muy interesante que nos comentara la evolución del paciente.

Atte, Gustavo Chiodetti, médico anestesiólogo.

Am J Emerg Med 1994 May;12(3):350-2 Related Articles, Books, LinkOut

Spinal epidural hematoma associated with cocaine abuse.

Huff JS. Department of Emergency Medicine, Eastern Virginia Graduate School

of Medicine, Norfolk 23507.

A 41-year-old man rapidly developed quadriparesis moments after injecting cocaine. Investigation discovered a cervical spinal epidural hematoma. This complication of cocaine abuse has not been previously reported.

Arch Neurol 1996 Aug;53(8):819-21 Related Articles, Books, LinkOut

Spontaneous spinal epidural hematoma. Another neurologic complication of

cocaine?

Samkoff LM, Daras M, Kleiman AR, Koppel BS.

Gustavo Chiodetti

Estiamdos Amigos:

¿La bradicardia de este paciente estará realmente relacionada con el consumo de drogas? Una posibilidad podría ser que la afección neurológica afecte la inervación simpática del corazón. Un dato interesante sería saber la duración del QT.

Marcelo Chambo

Neuquén Argentina

Prezado Dr Gustavo Chiodetti : o seu aporte me parecera muito interesante, mesmo sendo raro voce menciona "o hematoma epidural poderia explicar o quadro neurológico e a bradiarritmia por comprometimento do simpático cardiotorácico".

Justamente faz pouco tempo nos estavamos revisando um método nao invasivo que permite detectar o acometimento autonômico simpático.

Vejamos este raciocinio: se o hematoma é de localização epidural significa que pode comprometer o primeiro neuronio autonómico simpático, uma vez que o mesmo possui seu soma (corpo celular axónico) nas células que dão origem as fibras simpáticas pré-ganglionares que encontram-se localizadas na coluna intermedio-lateral de Stilling da medula espinal e seus axones (estes agredidos pelo hematoma) emergem acompanhando as raizes anteriores da medula para ir ao encontro do segundo neuronio autonômico simpático que tem seu corpo celular nos ganglios paravertebrais. De sorte que se há comprometimento deste primeiro neuronio necessariamente terá que existir menor aporte de neurotransmissor na sinapse pré-ganglionar do simpático que atua no coração, isto é, os tres ganglios cervicais (superior ou estrelado, médio e inferior) e os 4 a 6 primeiros segmentos torácicos.. Pois bem, utilizando uma técnica de neurotransmissores das imagens neuro-cardíacas que se constitue no único método não invasivo para visualizar os aspectos pré e pos sinápticos do sistema nervoso autónomo (SNA) permitindo examinar a integridade simpática, função e alterações regionais nos neuronios do SNA (Morozumi T, Kusuoka H, Fukuchi K, et al. Myocardial iodine-123-metaiodobenzylguanidine images and autonomic nerve activity in normal subjects. J Nucl Med 38:49-52, 1997).

O método tem sido denominado de várias maneiras, e usa como radiotrazador o chamado Iodine-123 metaiodobenzylguanidine (MIBG) que permite imagens de "single-photon emission tomography (SPECT) conhecidas como "123I-metaiodobenzylguanidine scintigraphy".

O isótopo metayodo-benzil guanidina (MIBG) foi inicialmente desenvolvido para o diagnóstico de feocromocitoma. Atualmente é usado nesta entidade quando a TC e a RNM falharam ou nos casos de tumores múltiples. Trata-se de um análogo da epinefrina e e da guanetidina, um falso neurotransmissor que possui captação, transporte, armazenamento e eliminação que imita a noradrenalina nas terminações pré-sinápicas do coração (upstake 1).

Por tanto o radiofármaco tem metabolismo muito semelhante a norepinefrina natural sendo captado dentro das células e armazenado em vesículas e nao é catabolizado. Adicionalmente é captado pelo tecido nervoso (upstake 2).

O clearenece do MIBG é de 5% a 12% em 3h a 4h e coincidente com o da noradrenalina. Estudos realizados com este composto tem demonstrado sua grande capacidade para delimitar denervação regional no coração.

As neuropatias generalizadas como a diabética se associam a marcada diminuição do upstake 1 do [123I]MIBG dentro do coração (Sisson JC, Shapiro B, Meyers L, Mallette S, Mangner TJ, Wieland DM, Glowniak JV, Sherman P, Beierwaltes WH. Metaiodobenzylguanidine to map scintigraphically the adrenergic nervous system in man. J Nucl Med 1987;28:1625-360).

Quais são as aplicações clínicas atuais desta técnica de imagens neuro-cardíacas ????

Resposta:

Várias são as entidades onde o método tem se mostrado útil sendo a aplicação mais usada como marcador de prognóstico não invasivo na insuficiência cardíaca.

1) **Estudo da insuficiência cardíaca**: Sabe-se que níveis elevados de norepinefrina estão associados a pior prognóstico. A ICC associa-se também a menor depósito de adrenalina no coração. Um nivel de atividade da MIBG anormalmente baixo associa-se a mau prognóstico independente da FE. A taxa de MIBG uptake (captação) expresa-se pela relação coração/mediastino (heart/mediastinal ratio) Se esta for menor do que 1.2 se associa a prognóstio ruim. (Cohen-Solal A, Esanu Y, Logeart D, et al. Cardiac metaiodobenzylguanidine uptake in patients with moderate chronic heart fairlure: Relationship with peak oxygen uptake and prognosis. J Am Coll Cardiol 1999; 33:759).

2) **Miocardiopatia hipertrófica** (Shimizu M, et al. Long term course and cardiac sympathetic nerve activity in patients with hypertrophic cardiomiopathy. Am Hear J 1992; 67: 155-160) (Chen SL, Uehara T, Morozumi T, et al. Myocardial metabolism of 123I-BMIPP in patients with hypertrophic cardiomyopathy: Assessment by radial long-axis SPET. Nucl Med Comm 16:336-343, 1995) (Nishimura T, Nagata S, Uehara T, et al. Prognosis of hypertrophic cardiomyopathy: Assessment by 123I-BMIPP (b-methyl-p-(123I) iodophenyl penta-alpha@decanoic acid) myocardial single photon emission computed tomography. Ann Nucl Med 10:71-78, 1996). (Ito Y, Hasegawa S, Yamaguchi H,

et al. Relation between thallium-201/iodine 123-BMIPP subtraction and fluorine 18 deoxyglucose polar maps in patients with hypertrophic cardiomyopathy. J Nucl Cardiol 7:16-22, 2000).

3) **Miocardiopatia dilatada idiopática** (Maeno M, et al: The significance of 201T123I MIGB (metaiodobenzylguanidine) mismatched myocardial regions for predicting ventricular tachycardia in patients with idiophatic dilated cardiomyopaty Kaku Iguaku 1993: 30:1221-29)

4) **Diabetes Mellitus**: quando a mesma tem neuropatia apresenta um MIBG uptake baixo. Esto explica o maior número de cardiopatia silente sem dor nesta entidade.

5) **Fase aguda do infarto**. Nesta fase observa-se menor MIBG uptake que mais tarde se recupera. (Haitikainen J, Kuikka J, Mantysaari M, et al. Sympathetic reinnervation after acute myocardial infarction. Am J Cardiol.1996; 77:5-9). (Shimonagata T, Ishida,Y, Hayashida K, et al. Scintigraphic assessment of silent myocardial ischaemia after early infarction using myocardial SPET imaging with 201Tl and 123I-MIBG. Nucl Med Comm 16:893-900, 1995). (Nishimura T, Nishimura S, Kajiya T, et. al. Prediction of functional recovery and prognosis in patients with acute myocardial infarction by 123I-BMIPP and 201Tl myocardial single photon emission computed tomography: A multicenter trial. Ann Nucl Med 12: 237-248, 1998).

6) **Coração Transplantado**: este revela menor MIBG uptake. que volta a aumentar com os sinais de reinervação (De Marco T, Dae M, Yuen-Green MS, et al. 123I-metaiodobenzylguanidine scintigraphic assesment of the transplanted human heart: Evidence for late reinnervation. J Am Coll Cardiol. 1995; 25:927-32.).

7) **Displasia arritmogênica do VD** (Witcher T, Hindricks G, Lerch H, et al. Regional myocardial sympathetic dysinnervation in arrhythmogenic right ventricular cardiomyopathy. An analysis using123I-metaiodobenzylguanidine scintigraphy. Circulation 1994; 89:667).

8) **Sindromes do QT longo**: A parede anterior do ventrículo esquerdo está inervada pelo simpático direito e nestes pacientes, possui uma repolarização ventricular mais demorada, em relação a parede ínfero-posterior inervada pelo simpático esquerdo. Esta não homogênea repolarização entre a parede anterior e a posterior seria parcialmente responsável pela manutenção da arritmia pelo mecanismo de reentrada aumentando a instabilidade miocárdica. A estimulação simpática como o estresse físico ou emocional ocasionam nos pacientes portadores de SQTL congênito:

1) aumento ainda maior do QTc,

2) frequente aparecimento de arritmias por ocasiões severas como as torsades de pointes (TdP),

3) aumento da dispersão transmural da repolarização como consequencia de encurtamento do PAT do epicardio, endocardio mais não das células M. O fenômeno é mais marcado na variedade LQT1 do que na LQT2 e LQT3. Esta última variedade alélica com a síndrome de Brugada é insensível as catecolaminas. Contrariamente a terapia antiadrenérgica com beta-bloqueadores ou a estelectomia esquerda melhora substancialmente o prognóstico. O último procedimento está indicado quando os beta-bloqueadores encontram-se contra-indicados ou quando mesmo com seu uso o paciente não se encontra totalmente protegido.

9) **Sindrome de Brugada (SB)** nesta entidade, Wichter et al. encontraram redução na captação de 123I-metaiodobenzylguanidine na parede ínfero-septal do VE quando comparados a sujeitos normais indicativa de disfunção pré-sináptica (Wichter T, Matheja P, Eckardt L, Kies P, Schafers K, Schulze-Bahr E, Haverkamp W, Borggrefe M, Schober O, Breithardt G, Schafers M. Cardiac autonomic dysfunction in Brugada syndrome. Circulation 2002;105:702-6).

Estes fatos já tinha sido detectados num paciente por Noda et al. (Nomura M, Nada T, Endo J, Kondo Y, Yukinaka M, Saito K, Ito S, Mori H, Nakaya Y, Shinomiya H. Brugada syndrome associated with an autonomic disorder. Heart 1998;80:194-6).

10) **Fibrilação Ventricular idiopática**: da mesma forma que na síndrome de Brugada Schafers et al verificaram redução na captação de 123I-metaiodobenzylguanidine na parede posterior do VE indicando disfunção pré-sináptica nesta entidade (Schafers M, Wichter T, Lerch H, et al. Cardiac 123I-MIBG uptake in idiopathic ventricular tachycardia and fibrillation. J Nucl Med 1999;40:1-5).

11) **Cardiotoxicidade pelo uso de quimioterápicos** (Valdes Olmos R, tem Bokkel Huinink WW, Greve J et al. I-123 MIBG and serial radionuclide angiocardiography in doxorubicin-related cardiotoxicity. Cin Nucl Med 1992; 17:163-70).

Finalmente em teoria poderiamos encontrar uma aplicação de número

12 no **hematoma epidural** mencionado.

No les parece coherente?

Grato.

Andrés R. Pérez Riera

Queridos amigos:

Sin animo de polemizar ni mucho menos con la excelente revisión de Andrés sobre el MIGB, como método incruento para investigar el compromiso del SNA y que propone como indicación nro 12 para diagnosticar este presunto hematoma epidural sospechado por Gustavo Chiodetti, me pregunto si para tal cometido, ¿no sería más útil y más económico una TAC o una RMN?

Me parece, -y perdón por incursionar en elucubraciones neuroquirúrgicas, que no me competen-, que si tal fuera la sospecha clínica, más que investigar el compromiso del SNA, habría que recurrir a un método que permita rápidamente tomar o rechazar la indicación quirúrgica, para liberar el canal medular de la compresión por el hematoma (si se descubriere que es el responsable)

Un abrazo

Edgardo Schapachnik

Prezado Edgardo: como ja comentaramos o método em várias circunstancias constitui em uma opçao a TC e a RNM. Esto foi expressado no meu comentário por exemplo no caso do feocromocitoma entidade que possui uma indicaçao cirúrgica clara. Neste caso em particular só se indica quando o feo responde a tumores múltiples os quais costumam estar associados a neoplasias endócrinas múltiplas MEN a qual apresenta dois tipos: Tipo 2A: ou síndrome de Sipple associado ao carcinoma medular da tiroides. Tipo 2B: ganglioneuromas mucosos.

O comentário que fizera é apenas um exercicio teórico, para eliminar a "ferrrugem" de nossos neuronios. Nunca li que tenha sido utilizado para ese fim o número "12". Apenas tenho a impressao que ao modificar o autónomo simpático pode alterar o" turnover" adrenérgico.

Recentemente estive revisando e encontrei esto que te da toda a raçao a RNC é o " padrão ouro " para o diagnóstico:

Vejam estes tres relatos de casos de literatura em homen de media idade (45a), idoso (77a) e criança (10a).

1) SPONTANEOUS RECOVERY OF SPINAL EPIDURAL HEMATOMA

H. Eskandary, MD; H. Reihani Kermani, MD

Spinal epidural hematomas are rare clinical entities that present with severe and acute pain followed by progressive motor and sensory deficit, and which have been associated with trauma, coagulopathies, arteriovenous malformation and neoplasm.1-3 In the absence of significant trauma or other discernible cause, they have been described as spontaneous.3-4 We report a case of spontaneous spinal epidural hematoma (SSEH) in a 45-year-old patient with typical presentation, diagnosed by magnetic resonance imaging (MRI) scan, who was managed conservatively in view of his rapid neurological improvement.

Case Report

A 45-year-old man admitted to our hospital had suddenly developed severe pain at the lower neck and upper thoracic spine, and became paraplegic a few days before admission. He had some pain relief and experienced improvement in the weakness in his legs. There was previous history of fecal and urinary dysfunction. Apart from that there was no past medical history of note, and he was on no medication. Examination revealed a fit normotensive man with no abnormalities in the chest or cardiovascular system. There was no sign of meningeal irritation. Examination of cranial nerves was unremarkable. Tone and muscle power were normal in the upper limbs, but decreased in the proximal and distal part of the lower limbs. The deep tendon reflexes were present with reinforcement in the upper limbs but were exaggerated in the lower limbs. There was sensory loss below the T2 bilaterally. Laboratory investigations including CBC, prothrombin time, partial thromboplastin time, and platelet count, were normal. Electrocardiogram was unremarkable and random blood glucose and electrolytes were normal, as well as chest and plain x-ray of the spine. MRI showed a space-occupying lesion in the posterior part of the spinal canal at the level of D1 and D2 vertebrae, which was hypointense in T1- and T2-weighted

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image (Figure 1). The sagittal MR view clearly demonstrated the epidural location of the lesion and the spinal cord compression toward the anterior aspect of the spinal canal.

On admission, the patient was found to have improved symptomatically. This was confirmed by a physical examination that showed paraparesis, and not complete paralysis as before. The patient's neurological state after admission, both subjectively and objectively, continued to improve dramatically. An operative approach was initially postponed, and ultimately abandoned. At follow-up two weeks later, the patient had no complaints, and he could stand and walk normally. On examination, his motor function and reflexes were normal, and he had no sensory deficit. An MRI two weeks later showed that the lesion had disappeared completely (figure 2).

Discussion

Mattel et al. found over 300 cases of spinal hematoma in the world literature up to 1987.5 Bruyn and Bosmu found 174 cases in the literature up to 1974. More than half the cases are spontaneous.3,4,6 The patients' age in the cases reported in the literature range from six months to 79 years, but SSEH is most frequently seen in the middle-aged or elderly.3,7 There is no significant sex difference in adults, but in children older than two years, males outnumber females.3,4,7 The majority of lesions occur in the thoracic region and extend into the cervicothoracic or thoracolumbar area.2,3,8,9 In a literature review of 155 cases of spinal epidural hematoma, Foo and Rossier concluded that the majority of cases (41%) were idiopathic.10 Although the majority of epidural hematomas are idiopathic, among the possible etiologies the most frequent is anticoagulant therapy (21%), followed by vascular malformation (6.8%). Trauma, vigorous physical exercise, poisoning with raticides, high blood pressure, neoplasms, vertebral angioma, acute infection and iatrogenic complications are less often causes.1-3,6,7,9 Until recently, epidural veins were considered the source of hemorrhage in spinal epidural hematoma because arteries in spinal epidural space were considered scarce.3 Some authors underline the possibility that the valveless vertebral venous plexus may not be able to withstand brusque elevation of pressure, concomitant even to physiologic activities such as FIGURE 1. MRI of a spontaneous spinal epidural hematoma. T1-weighted sagittal section of the spine (arrow). voiding, coughing and sneezing.4,11 The role of venous plexus structures in SSEH has been denied by other authors, who hypothesize an arterial origin of the lesion.4 Beatty and Winston suggested the spinal epidural arteries as the hemorrhage source.12 They based their view on the studies of Crock and Yoshizawu on the blood supply of the vertebral column and spinal cord, which showed a spinal epidural arterial network much more developed than previously thought, and on the fact that intraspinal venous pressure is inferior to intrathecal pressure, and so should not significantly compress the spinal cord in case of hemorrhage. Most spontaneous spinal epidural hematoma are classified as acute, resulting in paraplegia, quadriplegia and even death. The symptoms rarely have a slowly progressive course and a chronic type has been reported.1-6,10-13

Differential diagnosis of SSEH includes acute herniated intervertebral disc, acute ischemia of spinal cord, epidural tumor or abscess, spondylitis, transverse myelitis, hematomyelia, or even dissecting aortic aneurysm.3,9,13 MRI scan is the diagnostic procedure of choice.1,10 Although the prognosis of SSEH without surgery or with delayed surgery is poor, spontaneous resolution does occur and can result in clinical improvement.11 Emery

and Cochrane have reported dramatic reduction of pain and paralysis followed by myelography.10 Suzuki et al. summarized 22 cases of spontaneous remission without, or before surgery.14 Our case is an addition to those.

In a patient with spinal hematoma who does not recover from cord compression, a predominantly demyelinating lesion probably progresses into a

FIGURE 2. After two weeks, T1-weighted sagittal sections demonstrating the complete resolution of the epidural hematoma (arrow). predominantly axotomizing lesion, whereas a patient who recovers probably sustains only a demyelinating lesion.15 To date, the therapy of choice for spontaneous spinal epidural hematoma has been the prompt surgical evacuation of the hematoma by laminectomies, usually with favorable results. There has not been any role for conservative treatment in the management of this condition.

We have described a patient who had a spontaneous upper thoracic spinal epidural hematoma that resolved spontaneously and completely. The excellent outcome in this patient and several similar cases from the literature suggests that, in certain rare cases, surgical intervention is not mandatory. Thus, in the unusual case in which the patient's rapid neurological deterioration is followed by sustained neurological recovery, nonoperative therapy may be appropriate.

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2) Acute Spontaneous Spinal Epidural Hematoma

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REPORT OF A CASE

A 77-year-old woman was admitted to a general medical service with rapidly progressive asymmetric leg weakness. The neurocritical care service was immediately consulted, and a history of sudden onset of severe lower back pain immediately preceding the leg weakness was reported. On examination, the patient had an asymmetric flaccid paraparesis, sensory level at T12, and urinary retention. Magnetic resonance imaging (MRI) was performed 5 hours after the symptom onset. Figure 1 shows an area of hyperintensity (bright signal) on a sagittal T2-weighted MRI displacing the spinal cord and extending from T6-7 to L2-3. The areas of decreased signal intensity likely represent the presence of deoxyhemoglobin within the acute hematoma. The lesion markedly deforms the spinal cord, as shown in the axial T2-weighted images (Figure 2), and appears as an area of relative isointensity compared with the spinal cord on T1-weighted images (Figure 3). The MRI sequences are consistent with an acute hematoma. The hematoma was evacuated 11 hours following the symptom onset. Three weeks later, the patient was walking with a 4-pronged walker. No underlying cause, such as recent trauma, spinal catheterization, or coagulopathy, was identified.

COMMENT

Most cases of acute spontaneous spinal epidural hematoma occur in older patients and are likely caused by the rupture of an epidural venous plexus.1 The outcome may be linked to the location of the hematoma and the timing of an operation.2 The clinical suspicion of an acute spinal epidural hematoma is an indication for an emergent MRI of the spinal cord. The chronological characteristics of an MRI of a spinal epidural hematoma

are similar to those seen with intracranial hemorrhage,3 with some specific identifying features.4 In the hyperacute stage, the spinal epidural hematoma may appear as isointense on T1-weighted images and as isointense to mildly hyperintense on T2-weighted images as a result of the presence of intracellular oxyhemoglobin, although in practice, this stage is not commonly observed.4 The usual finding in the first 24 hours is the appearance of the hematoma as an area of predominantly T2-weighted hyperintensity in combination with focal areas of T2 hypointensity (caused by the presence of intracellular deoxyhemoglobin), with isointensity present on T1-weighted images. A conventional gradient-echo sequence may also be helpful, with acute blood appearing hyperintense on this sequence and structures containing cerebrospinal fluid, such as an epidermoid or arachnoid cyst, appearing hypointense.4 Magnetic resonance imaging performed after a few days (early subacute hematoma) will show the lesion as an area of T1-weighted hyperintensity and as T2-weighted hypointensity as a result of the presence of intracellular methemoglobin.

3) Case Report

A 10-year-old male presented with neck pain followed by sudden symmetrical onset of weakness involving all four limbs and accompanied by loss of all sensations over the trunk and limbs of one week duration. There was loss of bladder and bowel sensations with retention. There was no history of preceding febrile illness, recent intramuscular injections, local trauma or tuberculous contact. He was fully immunized. The child had received no treatment apart from symptomatic care at a local hospital prior to presentation.

On clinical evaluation, all vital parameters and anthropometry were normal. Neurologically, higher functions and cranial nerve examinations were normal. There was flaccid quadriplegia, deep tendon reflexes were exaggerated and plantar response was absent bilaterally. Sensory system examination revealed a loss of all modalities at and below C5 spinal dermatome. There was a band of hyperesthesia at C4 level, just below the clavicles. Sensations over the face and neck were preserved. Neck stiffness could be elicited on flexing, though there was no local tenderness over the cervical spine. All other systemic examination was normal.

Clinically, a diagnosis of acute transverse myelitis was considered and treatment with intravenous methyl prednisolone was begun, but the presence of neck stiffness prompted an early referral for MR imaging. MRI of the spine revealed an anterior epidural hematoma extending from C2 to C6 vertebral segments with its epicenter at C5 level and causing spinal cord compression (Figs. 1 & 2).

Fig. 1. MRI T1 weighted image of the cervicodorsal spine, saggital section, showing an anterior epidural hematoma (appearing as a linear hyperintense signal) extending from C2 to C6 vertebral level with its epicenter at C5 level and causing cord compression. Fig. 2. MRI_transverse section of the cervical spinal cord through C4-C5 junction showing the

anterior epidural hematoma (arrow) appearing white on the T2 weighted image. The cord shows an altered signal appearing as a hyperintensity due to edema/ischemia.

Subsequently the coagulation profile was discovered to be abnormal: Activated PTT was prolonged (patient_74 seconds, control_47 seconds) which was corrected on mixing (patient-49 seconds, control-47 seconds). The prothrombin time was normal (patient_16 seconds, control_14 seconds). Factor VIII assay revealed a level of less than 1%. Other routine hematological parameters were normal.

Retrospectively, on careful enquiry the boy denied history of indirect trauma. There were no complaints of easy bruising, hematomas after DPT vaccine, prolonged oozing from loose avulsed teeth or mucous membrane bleeds. One of the maternal uncles had been noted to develop recurrent swelling of the knee followed by crippling deformity, but had never sought medical advice.

The child was managed conservatively with infusions of factor VIII concentrate. There was some improvement by the end of 2 weeks, power had returned in the distal joints to 2/5 Other neurological signs had not changed. Repeat MRI after 12 weeks showed complete resolution of the hematoma. Neurologically, a flicker of movement could be elicited at proximal joints, but no appreciable improve-ment was noted distally. Anesthesia persisted and bladder sensations had not returned. Lack of complete recovery was attributed to delay in diagnosis and treatment.

Discussion

Spontaneous CNS bleed is an uncommon manifestation of hemophilia, and is usually precipitated by trauma, however trivial. Intraspinal bleeds account for 2-8% of CNS hemorrhages(2). Spinal epidural hematomas are rare; especially in children and exceptionally present as the first manifestation of severe hemophilia(3). The unusual feature of this case was the spontaneous nature of the epidural bleed and the complete absence of any manifestations prior to this event at the age of 10 years, despite severe factor VIII deficiency. Epidural hematomas are more common posterior to the spinal cord, anterior hematomas being relatively infrequent(4). The anterior location of the bleed in our child was the other unusual feature. Most of the cases reported have been infants or children less than 5 years of age(2,3,5-9), the youngest being 3 months old(7). Signs and symptoms are often atypical in infants, making the diagnosis difficult(2,3,6). In a literature review, less than 20 pediatric patients were reported with spinal hematomas(2,3,5-9) and none from India.

Surgical evacuation of hematomas by decompression laminectomy has its inherent complications and mortality as high as 22%(5). Recent reviews support treatment of selected cases with mild or stable neurological deficit conservatively with factor VIII concentrate infusions begun as early during the course as possible(2,5,6,8,9). Neurological recovery is gradual and complete recovery is possible over several weeks(2,5,6). However decompression laminectomy needs to be considered where diagnosis and treatment have been delayed and in cases where symptoms do not resolve with conservative management or are progressive(6,9).

This presentation stresses the need for prompt neuroimaging in every pediatric patient, especially a male, presenting with signs of acute transverse myelitis for accurate diagnosis of the underlying disorder so that specific treatment can prevent or reduce neurological sequel. Non-invasive modalities like CT or MRI are preferable(5,6,9).

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Queridos amigos:

El colega Roberto Balado hace llegar este aporte:

Estimado Edgardo y colegas suscriptos.

Leyendo los múltiples y excelentes aportes, quisiese agregar un comentario.

Me resulta más fácil de explicar la bradicardia y el cuadro neurológico pensando en el etilismo y la elevada frecuencia en que algunos adictos abusan de diferentes tóxicos, entre los cuales suele estar el alcohol en forma excesiva y descontrolada.

Agradecido por tu servicio

Roberto Balado