Aneurysmal bone cyst: A rare cause of medullary compression and painful scoliosis in children

Kyste osseux anévrismal : une cause rare de compression médullaire et de scoliose douloureuse chez l'enfant

Back pain with symptoms of spinal cord compression is not common in children and has a wide variety of causes [1], especially tumors. We present the case of a young patient with a history of back pain. He had neurological signs and scoliosis posture caused by an aneurysmal bone cyst (ABC) of the thoracic vertebrae. This case illustrates a less frequently recognized mechanism of spinal cord compression by such cysts.

Case

A 14-year-old boy with no history of trauma presented to the emergency department with painful thoracic scoliosis, which had begun two years earlier and steadily become worse, as well as with spastic paraparesis with intermittent urinary symptoms for two months. In view of this clinical picture of medullary compression, he underwent magnetic resonance imaging (MRI) of the spinal column (figure 1a and b), which showed a large expansile lesion from T7 to T11. It developed from the posterior arch of T9, extending intraspinally with compression of the spinal cord, to the left, and involvement of the right postero-lateral portion of the vertebral body (VB). This multilocular cystic lesion was seen as hypointense T1-weighted and hyperintense T2-weighted signals, showed fluid-fluid levels delineated by thin septa, and was significantly enhanced after contrast injection. The lesion also appeared to involve the dorsal soft tissue. These MRI findings were consistent with an ABC. The computerized tomography (CT) examination (figure 2a and b) showed the exact location and extent of bone destruction and the integrity of the cortical bone. Surgical resection of this lesion was performed and the spinal cord decompressed. Histopathological examination of the lesion confirmed an ABC (figure 3).

Discussion

Back pain is an uncommon symptom in children and has a wide variety of causes, including a number of tumors. ABCs account for 1.5–6% of benign bone tumors and most often (60% of cases) affect subjects younger than 20 years [2–4]. It is located on the spine in 6–20% of cases, in the dorsal segment in 34% of these [5]. The posterior elements and the pedicles are usually affected first, and in 60–70% of cases the lesion extends into the vertebral body [5,6]. The spinal topography of this benign tumor creates a serious neurological risk. Clinical findings of spinal ABC are usually nonspecific, such as diffuse pain accompanied by stiffness in the back. The initial presentation is due to neurological symptoms in 60–70% of cases [6]. A palpable mass or backache that is aggravated in the supine position alerts the physician to this problem. Depending on the level of involvement and the extent of spinal cord compression, a wide variety of neurological symptoms and signs may be noted later, ranging from mild radiculopathy to complete paraplegia or tetraplegia. Acute spinal cord compression can occur in the absence of vertebral body collapse if there is a break in the posterior cortex of the body that results in epidural extension of the lesion [7]. Other signs include scoliosis due to back pain and muscle spasm, paresthesia, and weakness. Scoliosis and kyphosis have been noted in roughly 10–15% of cases [5]. In the presence of spinal cord compression the radiologic assessment must start with the spine MRI, with and without contrast product, and should include a CT scan to assess the extent of the bone lesion.

Radiography showed a lytic eccentric lesion with swollen and reorganized contours. The cyst has clear margins and often contains nonspecific trabeculations, which result from periosteal bone formation. The presence of a fine osseous plate in the form of a shell indicates the slowly progressive nature of the lesion; it can, however, be missed. In that case, the CT scan will be required [8]. It assesses the extension of the expansile lytic osseous lesion and sometimes shows fluid-fluid levels with a sloping hyperdense component (40–100 H) and a less dense supernatant (10–60 H). Acquisition after 10 min lying down and analysis in a very narrow window are both recommended [9]. CT typically reveals a characteristic soap-bubble appearance, which represents a ballooning, multilocular lytic lesion. Thin walls of cortical bone result from erosion and expansion of the cortex. Common findings include pathologic fracture or partial (or even sometimes completely) vertebral body collapse. Preoperative CT imaging is helpful for assessing pedicle and vertebral body integrity in anticipation of instrumented fusion [5].

MRI is more sensitive than CT [9,10]. It shows the fluid levels in T1-weighted sequences, probably because of the methemoglobin. These levels tend to disappear in T2 weighting, because
Figure 1

a and b: Sagittal and axial MR images revealed an expansile lesion involving the posterior arch of T9 and right posterior vertebral body in heterogeneous hyperintense T2-weighted signals with fluid-fluid levels delineated by thin septa. Note the intraductal extension with marked spinal cord compression at T9 and a broad longitudinal extension.

the two components (serous fluid and clot) both have a very intense signal. Fluid-fluid levels are unspecific, however, and can be found in osteosarcomas, giant-cell tumors, and chondroblastomas [9,11]. The lesion contains partitions or cystic cavities shown clearly after gadolinium injection. MRI is the best imaging method for demonstrating multiple internal septa [12]. On sagittal and axial images it can clearly show the epidural extension of the mass and its compressive effect on the dural sac and thoracic spinal cord. In our patient, the preoperative MRI indicated an aggressive lesion with little reparative osteogenesis, contiguous vertebral involvement, and infiltration of the surrounding soft tissue. Arteriography can show peripheral hypervascularization, and arteriovenous shunts and can specify the feeder pedicles of the cyst for possible preoperative embolization [8,13]. The scintigraphy shows a primarily peripheral nonspecific uptake in 65% of the cases, also found in unicameral bone cysts, giant-cell tumors, and chondroblastomas [8,13].

The exact pathogenesis of ABCs is still uncertain. Some authors have suggested that it may be preceded by traumatic fractures or subperiosteal hematoma, and others that their formation results from a repair process. They are generally thought of as a secondary vascular phenomenon superimposed on a preexisting lesion, thought to initiate periosteal or intraosseous arteriovenous malformations. The resultant hemodynamic forces, generated by high-pressure vascular channels, can rapidly erode the osseous trabeculae into a cystic cavity. The associated reactive changes within the endosteum and periosteum incite accelerated osteoblastic and osteoclastic activities, causing rapid bone remodeling consistent with the hemodynamic forces and giving the “lesion a ballooned, thin-shelled, and multisepa soap-bubble appearance” [6]. According to current concepts, ABCs may occur in the bone as a solitary lesion or can be found in association with other tumors, including giant cell tumors, chondroblastomas, hemangiomas, osteoblastomas, and fibrous dysplasia, or in association with a malignant process such as telangiectatic osteosarcoma [4].

The differential diagnosis must rule out both other benign spine tumors and malignant tumors. In increasing order of frequency, bone tumors in children are eosinophilic granulomas, osteoblastomas, and osteoid osteoma [13]. Microscopically, the composition of the cyst may be dense and cellular, containing plump stromal cells, multinuclear giant cells, and thin-walled blood vessels, or be a preponderantly fibrous tissue with enlarging vascular spaces [4]. Conventional treatment has been directed at the surgical removal of either the entire lesion or as much of it as possible.
Recurrence is seen in 10–44% of cases, with 90% recurring within 2 years [10]. Appropriate treatment of an ABC requires recognizing that it has a specific pathophysiological origin. Identification of the pre-existing lesion, if possible, is essential. If no coexistent lesion is identified, lesions are usually treated with curettage and bone grafting, with more aggressive treatment reserved for large lesions, or those compressing the spinal cord or nerve roots, and for recurrences. Treatment must be directed toward the most aggressive component present. Stabilization with osteosynthesis is usually performed next. Nevertheless, in young patients it is important to preserve the alignment of the spine and permit harmonious development until growth terminates [14].

Recently, embolotherapy has been used to treat vascular bone tumors to limit blood loss at surgery or as definitive therapy when surgery is not feasible. The goal of embolization is occlusion of the lesion's vascular supply without interfering with the vascularity of surrounding tissue or structures. Successful embolization of an ABC will result in progressive ossification within 2–4 months of the initial embolization, almost always beginning at the margins. Complete ossification may require 8–12 months or longer. The effect of embolotherapy is therefore unpredictable. Moreover, potential complications include the occlusion of normal adjacent tissue, iatrogenic pulmonary emboli and catheter-related complications. We did not use embolotherapy, due to the danger of interference with the blood supply to the spinal cord in case of a abrupt myelopathy within a critical collapse in the cervicothoracic region. The addition of postoperative radiotherapy not exceeding 2000 rads has been proposed if some infiltrative tissue remains after partial excision of a huge tumor. Radiation-induced complications include growth plate disturbance, myelopathy, gonadal damage, and sarcomatous changes. Intralesional injection of thrombogenic agents (Ethibloc®, or alcohol) and calcitonin into spinal ABCs has been reported, but further clinical experience is necessary to establish their efficacy [7].

*Figure 2*

a and b: Axial CT images showed a variable-density multilocular lytic lesion arising from the posterior arch vertebral body of T9 with a characteristic soap-bubble appearance (a)

*Figure 3*

Photomicrograph (HES × 100) of the tumor showing fragments of bone tissue with cystic space walled by fibrous or fibro-osseous tissue. Areas of bleeding, aggregation of multinucleated giant cells, fibrosis, chronic inflammatory cell infiltration, and granulation tissue are also seen.
Clinical cases

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References


Conclusion

Here we present a case of a childhood ABC with medullary compression and a scoliosis posture. ABCs of the spine present challenges due to their highly vascular nature and the risk of spinal destabilization with resultant neurological compromise. The potential for acute nerve root or spinal cord compression due to compromised bone integrity necessitates their aggressive management. The radiologic findings, especially MRI and CT-guided biopsy, can often be helpful in firmly establishing a diagnosis of ABC.

Conflicts of interest : None.

Anémic hémolytique auto-immune : un mode de révélation de l’infection par le virus de l’immunodéficience humaine

Autoimmune hemolytic anemia: An unusual revelation of human immunodeficiency virus

L’anémie hémolytique auto-immune (AHAI) complique rarement l’infection par le virus de l’immunodéficience humaine [1]. Cependant, elle peut être grave et engager le pronostic vital.

Les cas révélant cette infection sont encore plus rares [1]. Nous rapportons une observation originale par le caractère révélateur de l’AHAI d’une infection par le VIH chez une jeune femme.

Observation

Mme M. A., âgée de 36 ans, était admise pour prise en charge d’un syndrome anémique sévère d’installation rapide. L’examen clinique trouvait une pâleur cutanéomuqueuse franche avec un ictère généralisé et une splénomégalie à 3 travers de doigts, sans fièvre. L’hémogramme montrait un taux d’hémoglobine de 4,1 g/dL, un volume globulaire moyen (VGM) à 80 mm3 et une lymphopénie à 620 éléments/mm3. Les taux de LDH et de la bilirubine indirecte étaient augmentés (respectivement 2190 UI/L et 115 mg/L) et le taux d’haptoglobine effondré à 0,06 g/L. Le frottis sanguin objectivait la présence de sphérocytes. Le test de Coombs direct était positif et le typage d’auto-antigens a objectivé des anticorps chauds de type IgG ; la concentration corpusculaire moyenne en hémoglobine (CCMH) était à 32 %, un taux de globules rouges à 2,19 x 1012 éléments/mm3 et un taux de réticulocytes à 20 000 éléments/mm3. Il existait également une polynucléose neutrophile à 11 300 éléments/mm3 et une lymphopénie à 620 éléments/mm3. Les taux de LDH et de la bilirubine indirecte étaient augmentés (respectivement 2190 UI/L et 115 mg/L) et le taux d’haptoglobine effondré à 0,06 g/L. Le frottis sanguin objectivait la présence de sphérocytes. Le test de Coombs direct était positif et le typage d’auto-antigenes a objectivé des anticorps chauds de type IgG ; la