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Original Research Paper

Surgery

CASE REPORT: ATYPICAL VERTEBRAL HEMANGIOMA IN MULTIPLE HEREDITARY EXOSTOSES PATIENT

Sami Al Eissa	Assistant Professor, Consultant spine surgery Head of Orthopedic Surgery Division King Saud bin Abdulaziz University for Health Sciences (KSAU-HS) King Abdulaziz Medical City (KFH-KAMC) Riyadh- Saudi Arabia
Mohammed AlSalman	Assistant Professor, Consultant musculoskeletal radiology Radiology department King Saud bin Abdulaziz University for Health Sciences (KSAU-HS) King Abdulaziz Medical City (KFH-KAMC) Riyadh- Saudi Arabia
Majed S. Abaalkhail	Assistant consultant orthopedic surgery Orthopedic Surgery Division King Abdulaziz Medical City (KFH-KAMC) Riyadh- Saudi Arabia
Mohammed Hassan Alshehri	Assistant consultant orthopedic surgery Orthopedic Surgery Division King Abdulaziz Medical City (KFH-KAMC) Riyadh- Saudi Arabia
Adeeb Rashid Almohimeed	Musculoskeletal radiology fellow Radiology department King Abdulaziz Medical City (KFH-KAMC)Riyadh- Saudi Arabia
Mohammed Benmeakel*	MB, BS.Orthopedic resident, orthopedic surgery division King Fahad Hospital – King Abdulaziz Medical City (KFH-KAMC) Riyadh- Saudi Arabia *Corresponding Author

ABSTRACT Hereditary multiple exotoses patients are reported to have multiple osteochondromas that can present in the axial skeleton. Another entity of spine masses and lesions is Vertebral hemangiomas, which are well known for asymptomatic presentation and are classified as the most common begin lesions in the vertebral body. No case was reported in literature and showed an association between HME and atypical hemangioma. We are presenting a 27-year-old man known case of HME, with a cord compression picture, and a pathological diagnosis of atypical intraosseous hemangioma. His MRI finding suggested an atypical mass with malignant features in T8-T9 level. Our report is shining a light on decreasing the threshold of suspicion when dealing with HME patients who are presenting with atypical back pain; and we recommend evaluating them thoroughly.

KEYWORDS : vertebral hemangioma, exostoses, osteochondroma

INTRODUCTION

Atypical vertebral hemangiomas are usually described if they were symptomatic or they were located in unusual sites other than the vertebra body. They also can be categorized as unusual, based on their radiological or pathological appearance. However, benign hemangioma is the most common form of benign mass in the axial skeleton, and it was found in 11% of spine autopsies (1). Spine osteochondromas were rarely reported in literature, either in their presence as solitary mass or in the form of hereditary multiple exostoses (2). They located usually in long bones and mostly present as asymptomatic masses, although, HME presents in a familial pattern and as painful masses, causing deformity early in life. To our knowledge, there has not been any case in literature presenting a radiological finding of atypical osteocondroma in a hereditary multiple exostoses patient with pathological diagnosis of atypical hemangioma of the spine.

CASE REPORT

We had a 27-year-old gentleman, know case of hereditary multiple exostoses, presented to orthopedic clinic with right thigh mass, bilateral legs pain and anterior thighs accompanied by numbness in the left side more than the right. Based on this presentation his orthopedic surgeon to decide requesting lumbar MRI. The lumbar spine MRI showed L4-L5 herniated nucleus pulposus. He was referred to spine clinic at that time We noticed that he also started to show gait changes and complaining of frequent falls. On exam at that time he had power of 5 at the lower limbs and signs of upper motor neuron lesion. Then, he was sent for cervical and thoracic spine MRI to complete his evaluation. After all images were complete and 2 days before his follow up appointment, he presented to our emergency department with bilateral lower limbs acute progressive weakness (power 3 out of 5).

Whole spine MRI was done one week before this presentation (Figure 1 and 2); it was showing an abnormal T2 hyperintensity with marrow expansion and enhancement involving the left transverse process of T9 with extension to the same level pedicle and associated with epidural component at T8-T9 level, it was causing severe cord compression. Based on musculoskeletal radiologist review, it was interpreted as an atypical intraspinal osteochondroma with worrisome malignant features. Our pre-operative presumption was secondary chondrosarcoma with possibility of other malignant tumors.

This patient underwent emergent decompression and tumor debulking at T8-T9 level with posterior instrumentation from T6 to T11. Intra-operatively, we faced extensive bleeding while debulking the tumor. One week later, the pathological report showed an intraosseous hemangioma.

The patient post-operative course was extensive physiotherapy for ambulation; to be started gradually and as tolerated by the patient. At 3 months interval he had a follow up spine MRI, showing no residual extradural component of the tumor. He was followed for 14 months, and his motor status improved to power 5 out of 5; no recurrence was noticed. Although, the patient has had a right lower leg numbness and was diagnosed later with large osteochondroma at the posterior aspect of the proximal femur.

DISCUSSION

Hereditary multiple exostoses is a genetic autosomal dominant disorder. It was identified as a cause of cervical myelopathy in Yakkanti's review (5) of spinal osteochondromas (3). Zaijun et al (4) mention that most osteochondromas present in the posterior arch, however, Upadhyayain's review alerts us to symptomatic locations of osteochondromas e.g. spinal pedicle or lamina. In the scope of solitary vertebral osteochondroma, cervical spine is the most common site; previously, C1 had the majority of osetocondromas; while more recent reports showed that C2 and C3 are the most common sites of osteochondromas (6-8). This was followed by thoracic and lumbar spine equally.

Many atypical presentations of osteochondromas in the spine were reported; one of which is a case reported by Ko Ikuta and his colleagues (9), they diagnosed young gentleman with C5 osteochondroma araising from the lamina and same segment disc hereniation. Mesfin et al (10) reported one patient, known case of HME, with L5 mass that has transformed to secondary chondrosarcoma. This patient was completely asymptomatic except for palpable mass at the lower spine. Also, one case was reported in literature, showed young lady diagnosed previously with HME and presented with Brown Sequard Syndrome due to T3-T4 osteochondroma (11).

Upadhyayain and his colleagues (5) described 27 cases of HME with thoracic vertebral exostoses, 21 cases presented with neurological deficit; and all the 27 cases were treated surgically except for two that were conservatively dealt with. In regard to outcome, 5 cases had partial improvement or worse status after surgical intervention, although, one case was misdiagnosed as tuberculosis. In 1964, Solomon and his colleagues (12) reported many osteochondromas located in the spine, in HME patients, and all of them were symptomatic.

Bellasri et al (13) reported one case presented with neurological deficit caused by hemangioma extending to the spinal canal. That patient was a middle age lady with radiological findings of atypical hemangioma, It was located in T7 spinous process with extension to the lamina and causing narrowing of the spinal canal. Boriani and his colleagues (14) classified spine masses into 4 categories; Bellasri's case report was an example of type VI.

McEvoy et al (1) in reviewed atypical hemangiomas thoroughly and showed that they tend usually to present as aggressive masses since their histological appearance show more vascular structure and less fat presence. The extension of such masses into the extradural space was a sign of more aggressive behavior, which lead them to be symptomatic painful masses. However, none of their cases were known to have hereditary multiple exostoses.

Vertebral hemangiomas are mostly benign and have a charachtristic appearance on MRI; they usually present bright (increased intensity) on T1 and T2 weighted MRI images. In T1 they are bright due to their high fat content, while, in T2 images, they are bright due to high water composition (15). However, the atypical aggressive type of hemangioma has low intensity on T1 images; the reason for that is the reduction of fat contents intralesionally (16).

Treatment of all masses causing neurological deficit due to cord compression is mostly surgical, regardless of the cause. In most case reports of spine hemangiomas or osteocondromas that were causing narrowing of the spinal canal, they had a favorable prognosis after surgical intervention when cord decompression was the goal. Our case was unique by its unusual association of HME and atypical hemangioma. We recommend to fully evaluate all pateints with HME and to be aware of their symptomatic spine disorders; and to pick up all cases before they develop cord compression and neurological deficits.

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CONFLICT OF INTEREST

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FIGURES:

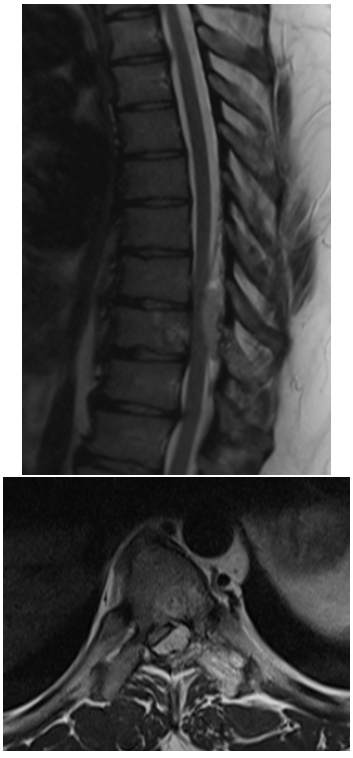

Figure 1a and 1b: T2 MRI images showing T8-T9 vertebral mass with appearance of atypical osteochondroma, causing severe cord compression in sagittal and axial cuts respectively.

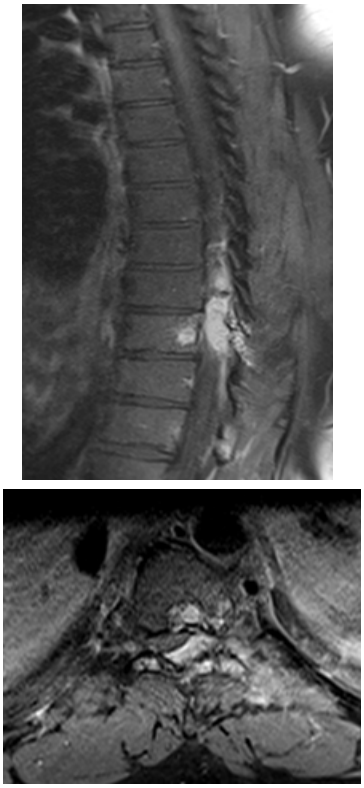

Figure 2a and 2b: MRI images with gadolinium contrast showing the T8-T9 vertebral mass in sagittal and axial cuts respectively.

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