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Case Report

Olliers Disease

Rib Head Chondrosarcoma associated with Olliers Disease – A Rare Differential for Paraparesis

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Olliers disease is a non-familial condition affecting 1:100000 (1,2). It is characterized by the presence of multiple enchondromas affecting small tubular bones of the hands, feet and other tubular long bones and flat bones. Malignant transformation of these enchondromas is as high as 5 – 50% (1,2). While cases of de novo chondrosarcomas involving the vertebral body pedicles causing cord compression and neurological deficit have been well documented. We present a rare case of Olliers disease with chondrosarcoma of the rib head who presented with paraparesis secondary to extradural cord compression with intracanalicular extension.

Keywords: Olliers Disease, Chondrosarcoma, Enchondromas, Chondrosarcomas

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Introduction

Olliers disease is a non-familial condition affecting 1:100000 (1,2). It is characterized by the presence of multiple enchondromas affecting small tubular bones of the hands, feet and other tubular long bones and flat bones.

Malignant transformation of these enchondromas is as high as 5 – 50% (1,2). While cases of de novo chondrosarcomas involving the vertebral body pedicles causing cord compression and neurological deficit have been well documented. We present a rare case of Olliers disease with chondrosarcoma of the rib head who presented with paraparesis secondary to extradural cord compression with intracanalicular extension.

Case Report

A 35 year-old gentleman presented to us with complaints of progressive weakness of the lower limbs ad difficulty in walking since 2 months and urinary retention since 15 days.

Neurological examination revealed and increased tone in both lower limbs (Modified Ashworth Grade 3) with distal muscles weaker than proximal (Gr 2/5 and Gr 3/5 respectively). He also had diminished sensation below D9 on the right side and D10 on the left side. General examination revealed multiple bony swellings over the long bones (bilateral femur, humerus and ulna) along with shortening of the right lower limb and a solitary large paravertebral swelling involving D6-10 ribs. The patient also had previous surgical scars over the left lateral chest wall secondary to surgeries he had for excision of similar bony lesions. Further review of his old case files revealed an excision of the lower ribs with intrathoracic and intra-abdominal extension 7 years ago. Pathology rendered a diagnosis of low-grade chondrosarcoma. He had a recurrence of the lesion 2 years postoperatively and had a resurgery done, however he underwent no radiation or adjuvant therapy.

Initial X-ray imaging showed significant scoliosis with chondroid mineralization of the 6th to 10th ribs on the left side. MRI of the dorsal spine revealed a dumbbell shaped lesion isointense on T1W and hyperintense on T2W with contrast enhancement (Figure 1). The lesion appeared to arise from the posterior end of D6 to D10 ribs with extension Through the D7-8 and D8-9 neural foramina into the extradural space with significant cord compression.

The lesion also showed a large intrathoracic and intraabdominal component.

Operative procedure: The patient was taken up for a D7-9 laminoplasty with gross total excision of the intracanalicular extradural component of the lesion. A CTVS and General surgery consult was sought for decompressing the intrathoracic and intra-abdominal component. Postoperatively the patient showed significant neurological and radiological (Figure 2) improvement with improvement in the motor deficits and bladder function returned to normal. Final histopathology reported it to be a low-grade chondrosarcoma.

Discussion

Olliers disease is a non-familial disease characterized by multiple enchondromas over the tubular bones of limbs and flat bones (1,2). The lesions are typically asymmetric with one side being usually predominantly involved however in our patient the lesions were distributed equally bilaterally.

Enchondromas typically affect the metaphysis and progressively involve the diaphysis (1). Verdegaal et al have categorized patient in to 3 groups. The first group of patients have enchondromas affecting short tubular bones of hands and feet, the second group have enchondromas affecting long tubular bones and flat bones while the third group is a combination of the first and second group. As per the classification our patient seemed to be in Group 2.

The conversion of enchondromas to their malignant variant is a well-known entity and depends on the age and duration of the lesion (1,2). As per the Verdegaal classification, Group 2 and Group 3 patients have a higher risk of malignant conversion (45% – 46%) whereas Group1 have only a 15% conversion rate. Pelvic enchondromas, in particular, have a high risk of malignant transformation. All central cartilaginous lesions should be considered malignant unless proved otherwise.

Patients with Olliers disease may have malignant transformation occurring at multiple sites simultaneously.

They may be synchronous or metachronous, as in our patient. In tubular bones, the presence of pain, sudden increase in size and large lobulated lesions of size > 5cms should arouse the suspicion of malignant conversion (3,4).

Radiologically, enchondromas are differentiated from solitary low-grade chondorsarcomas by the absence of cortical destruction and soft tissue extension however patients with Olliers disease have enchodromas that are locally aggressive and show cortical erosion and soft tissue extension. These findings may be seen even in cases without sarcomatous transformation (7). In addition a high degree of inter observer variability has also been reported (4). Cortical and soft tissue extension warrants investigation with Gadolinium enhanced MRI (Magnetic resonance imaging) and a biopsy should be performed in such cases. All patients with Olliers disease should routinely be screened for haemangiomas to rule out the possibility of Maffucci's syndrome. These patients have a higher incidence of malignant conversion and also have an increased incidence of hepatic and pancreatic malignancies, gliomas and acute myeloid leukaemias. While a cerebral and abdominal computed tomographic scan (CT) is warranted even with mild neurological or abdominal complaints, several authors recommend a routine screening in all cases of Maffucci's syndrome.

Chondrosarcomas associated with Olliers disease affecting the rib heads are well known as are primary enchondromas involving the vertebral bosdy and posterior elements and subsequently causing cord compression and neurological deficit (4,6).

The patient described in our report is the first documented case describing a rib head chondrosarcoma associated with Olliers disease causing neurological deficit secondary to intracanalicular extension and extradural cord compression. A study of solitary chondrosarcoma by Florenza et al reported a 30% and 37% mortality rate at 10 and 15 years respectively (8).

Verdegaal et al reported a 6% mortality in patients with chondrosarcomas associated with Olliers disease and in the 161 cases of Olliers disease and Maffucci's syndrome he reviewed, there was a 6% mortality secondary to chondrosarcomatous change of the lesions.

Surgical en bloc resection provides the best prognosis with tumor recurrence being about 25% in cases with tumor free margins (6). While some studies have proposed curettage with or without adjuvant therapy (phenol, bone cement, cyrotherapy) in Grade 1 chondrosarcomas of long bones (6), the results following partial excision have been unsatisfactory with poor prognosis.

In our patient total clearance of the intracanalicular and intraspinal component was possible leading to neurological recovery however due to extensive encroachment of abdominal and thoracic organs and vessel only a partial excision of the remaining tumor was possible.

Postoperatively, proton beam radiation was offered to the patient however multiple studies have shown no benefit with concurrent radiotherapy (4).

Figure Legends

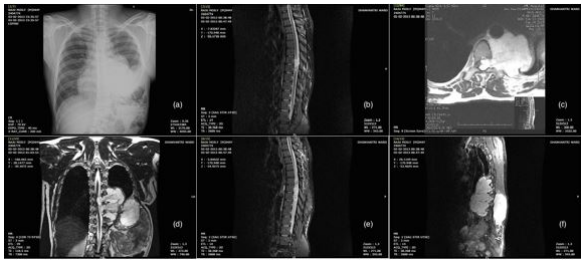

Figure 1: (a) Chest X Ray showing mineralization of the vertebrae with (b) , (c) and (d) showing a saggital, axial and coronal section showing a large well defined lobulated lesion which is T2W hyperintense, T1W iso-hypointense and peripherally enhancing on contrast arising from the posterior ends of left ribs extending from level of D6 to D 10 vertebrae. (d) The lesion is seen anteromedially encasing the aorta and abutting the pericardium and anterolaterally the lesion is seen displacing the left lower lobe with underlying collapse.

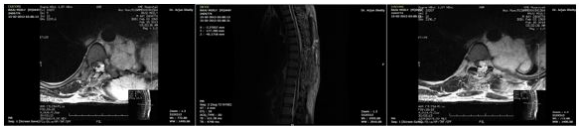

Figure 2: MRI LS spine (a) axial and (b) saggital sections showing absence of spinous processes of D6-D9 T2 and STIR hyperintensity showing mild contrast enhancement seen in the paraspinal region s/o post laminectomy status with inflammatory changes.

Widening of the left D 7-8 and D8-9 neural foraminae seen with extension of the lesion into the extra dural space. The epidural component and compression of the cord is reduced in the present scan compared with the previous scan s/o post op excision of the extradural component.

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