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EXTRA-ABDOMINAL DESMOID TUMOUR- A RARE PRESENTATION

Orthopaedics

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KEYWORDS

INTRODUCTION

Desmoid-type fibromatosis (DF) is defined by the World Health Organization (WHO) as a locally aggressive fibroblastic neoplasm arising usually in deep soft tissues with an infiltrative growth. These tumours have a tendency for local recurrence but lacks the metastatic potential that is expected of such aggressive lesions. It usually originates as a solitary tumour from the connective tissues; seldom multiple lesions can be isolated. These are usually intra-abdominal in location, but rarely occur in the extremities (extra-abdominal Desmoid). These lesions are rare, corresponding to less than 0.03% of tumors and 3% of all soft tissue tumors (1). Majority of cases occur between the ages of 15-60 year with peak incidence 25-35 years (2). Here, we present a rare case of an Extra-abdominal Desmoid tumour, otherwise called *aggressive fibromatosis* and its management in a 24yr old male patient.

CASE REPORT

A 24-year-old male who came with swelling of right hip for 6 months, pain over right hip for 1 month associated with difficulty in sitting, walking and squatting. This was not associated with any constitutional symptoms such as fever. On examination, 10 x15 cm swelling noted over the right gluteal region extending over the lateral aspect of the thigh with the overlying skin normal with a smooth surface. There was wasting noted of the musculature of the affected limb. On palpation, there was no local raise in temperature or tenderness noted over the swelling. A 12 x 20 cm swelling over the right gluteal region extending to lateral aspect of the thigh and inferior gluteal margin inferiorly, which was firm in consistency. The surface was irregular; margin merges with the surrounding muscle. An X-ray of bilateral hip and pelvis was done, which revealed a scalloping lesion in the ileum due to mass effect (Figure 1). MRI right hip and pelvis was done which revealed a soft tissue mass, appearing to penetrate the iliac wing outer cortex with surrounding pressure effects. HRCT Thorax was done to rule out pulmonary involvement, and there was no evidence of metastases.

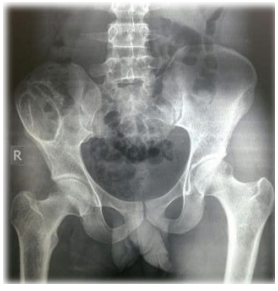

Figure 1: - X-ray of bilateral hip and pelvis

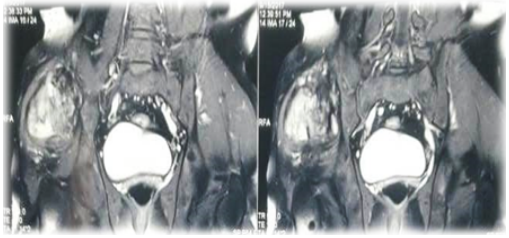

Figure 2: - MRI of the right hip and pelvis

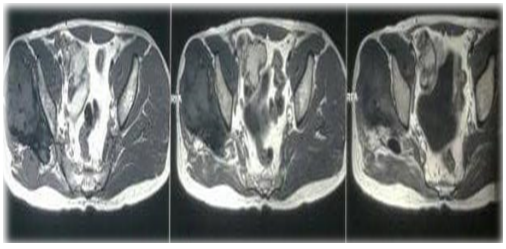

Figure 3: - MRI of right hip and pelvis

Open biopsy was done and reports showed feature suggestive of Desmoplastic fibroma of iliac crest. Patient was planned for tumour resection +/- pelvic reconstruction. Intra-operatively, the tumour was found to be within the bulk of the Gluteus maximus muscle, involving the nerve sheath of Sciatic nerve and there was no penetration of the tumor into the ileum. The entire tumor was debulked preserving the important structures (figure 4).

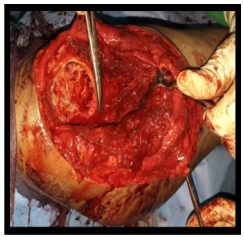

Figure 4: - Intra-operative findings where tumor was resected in toto

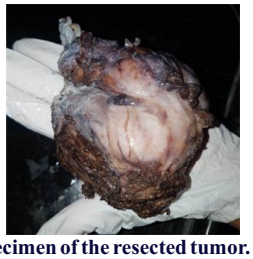

Figure 5- Gross Specimen of the resected tumor.

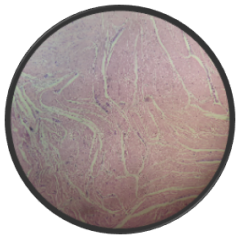

Figure 6: - microscopy showing spindle cells resembling fibroblasts with elongated nuclei

Histopathology confirmed the diagnosis of extra-abdominal desmoid type fibromatosis (figure 6).

Post-operatively, patient received adjuvant External Beam Radiotherapy 54Gy/27#/51/2 week. Patient was on regular follow-up

and symptom free with no disability.

Figure 7: - Post Operative follow-up showing functional range of movements

DISCUSSION

Extra-abdominal desmoid tumours, otherwise referred to as aggressive fibromatosis, are a rare occurrence in clinical practice. It often presents as a painless swelling; maybe associated with pain and motor weakness if the deeper structures such as muscles or nerves are involved. These tumours have a strong genetic preponderance such as Gardner's syndrome (in about 15% cases), pregnancy and female sex steroidogenic conditions (3). Hence it is found that extra-abdominal desmoid tumours have a female preponderance. Previous surgery, trauma, and hormonal imbalance have all been suggested as possible risk factors for the development of extra-abdominal desmoid lesions.

As these tumours are benign in nature, they can be managed by 'wait and see' approach. However, due to the large size causing compressive effects on the surrounding structures, surgery may be necessary. However, even with negative resection margins, it is important to note the high recurrence rate of 15–77% (4, 5) in such cases, obtaining a tumor free margin should not be the main treatment strategy. There are several adjuvant therapies, such as chemotherapy and radiotherapy. In this case, patient underwent debulking of the tumor with preservation of the sciatic nerve and post-operative Radiotherapy. In certain cases, it has been recommended that Chemotherapy with Vincristine and Methotrexate can be administered. Recent studies have shown that Doxorubicin-based treatment regimens have been used in the treatment of desmoid lesions and are often used in combination with Dacarbazine (6).

CONCLUSION

Extra-abdominal Desmoid tumours are a rare occurrence, which are benign yet locally aggressive lesions. These require a multi-disciplinary approach to treatment. With tumor debulking and post-operative appropriate adjuvant therapy, good clinical outcomes can be achieved.

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