COCCYGEAL CHORDOMA WITH INGUINAL LYMPH NODE AND SKIN METASTASIS -A CASE REPORT

Patil Rashmi K[1], Ameet G[2]

ABSTRACT

A 65 five year old man developed back pain associated with a coccygeal mass 2 years prior to presentation. Fine needle aspiration and excision biopsy of the coccygeal mass showed features of Chordoma. The ImmunoHistoChemical stains S-100, cytokeratin, epihelial membrane antigen, brachyury were all positive. After 15months of follow up patient developed local recurrence of tumour and left inguinal lymph node metastasis. Subsequently he developed cutaneous metastasis. The patient died 24 months later from complication of metastatic disease. Chordomas are rare, presenting only 1%-4% of malignant bone tumours, most often occur in the sacrococcygeal area. Metastasis to skin and lymph node is unusual. We report a rare case of coccygeal Chordoma with inguinal lymph node and skin metastasis.

Key words: Chordoma, coccygeal, lymph node, skin, metastasis

INTRODUCTION

Chordoma as a rare slow growing malignant tumour, arising from remnants of the foetal notochord accounts for 1%-4% of primary bone tumors. [1]They are generally considered locally aggressive, rarely if ever metastasizing tumours. The reported rates of metastases range from 10% to 48% [2, 3], in sacrococcygeal chordoma. The incidence of lymph node and skin metastasis is rare and is reported in few cases. [1, 2, 5, 9] FNAC has been found to be a useful tool in the preoperative diagnosis of chordoma. Moreover a combination of Histopahtological study and ImmunoHistoChemical study increases the accuracy of diagnosis.

The present case is described because of rarity of this tumour compounded with its infrequent inguinal lymph node and skin metastasis.

CASE REPORT

A 65 year old man presented with complaints of pain and swelling in the lower back region of 6 months duration. Examination revealed averagely built elderly male with mild pallor. A 4x2 cms soft, nonmobile mass was palpable in the sacrococcygeal region.

MRI showed a well defined soft tissue lesion below the sacrum eroding the coccyx. (Figure1A). Fine needle aspiration of coccygeal mass was done and stained with Giemsa and Papanicoloau stains.

Subsequently the patient was taken up for total surgical resection of the mass. The gross specimen consisted of single, soft, haemorrhagic tissue measuring 5x3x3.5 cms. (Figure2A) Sections were submitted for routine processing and staining with haematoxylin and eosin.

Diagnosis of chordoma was done on fine needle aspiration. (Figure1B). Haematoxylin and eosin stained tissue sections showed features of conventional Chordoma (Figure2B). ImmunoHistoChemical studies showed positivity for cytokeratin (CK), epithelial membrane antigen (EMA), Vimentin, S-100 and brachyury.

Follow up care- After 15 months of follow up, patient developed local recurrence and left inguinal lymphadenopathy. NMR scan of pelvis showed a recurrent lesion in the coccygeal region. Histological examination of resected, recurrent coccygeal lesion revealed conventional chordoma with abundant areas of necrosis. (Figure 3.A, 3 B) Fine needle aspirate of the left inguinal lymph nodes revealed metastatic deposits of chordoma. (Figure 4.A) Postoperatively radiotherapy was given to the patient for 1 month. Subsequently patient developed skin lesions, blue grey nodules involving chest and lower abdomen. (Figure 4.B) Skin lesion also showed features of metastatic chordoma. Patient died after 1year due to complications of metastasis.

[1]Assistant Professor, Department of Pathology, Belgaum Institute of medical sciences, Belgaum-590001 Karnataka Email: rashminimbal@rediffmail.com
[2]Assistant Professor, Department of Pathology, Belgaum Institute of medical sciences, Belgaum-590001, Karnataka, Email: ameet.bbo.belgaum@gmail.com

Figure1. A. Sagittal MRI scan showing well defined lesion involving the coccyx.

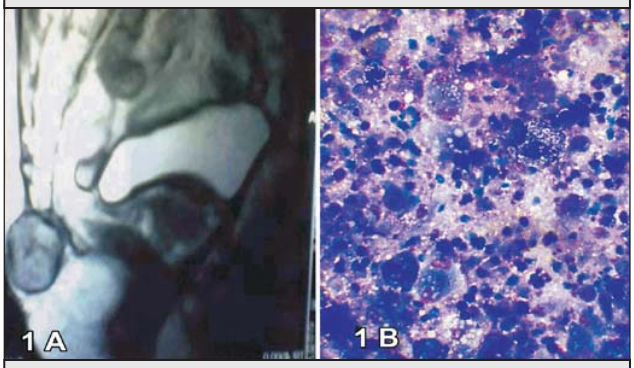

Figure 1.B. Microphotograph showing cytological appearance of chordoma –physaliphorous cells with vacuolated 'bubbly' cytoplasm (Giemsa X 400).

Figure.2.A. Photograph of fresh gross specimen of chordoma of coccyx showing red tan, haemorrhagic mass with glistening surface.

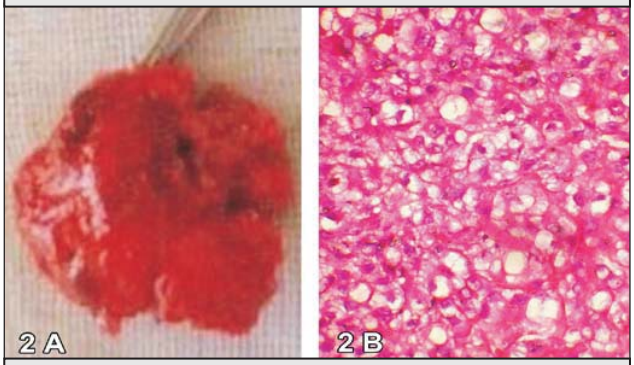

Figure.2.B. Microphotograph of conventional chordoma showing tumour cells arranged in cords surrounded by myxoid matrix with foci of necrosis (H&E, X400)

Figure 3.A. Microphotograph showing conventional chordoma with the diagnostic features of lobulations, myxoid matrix and physaliphorous cells in a cord arrangement (H&E, X 200).

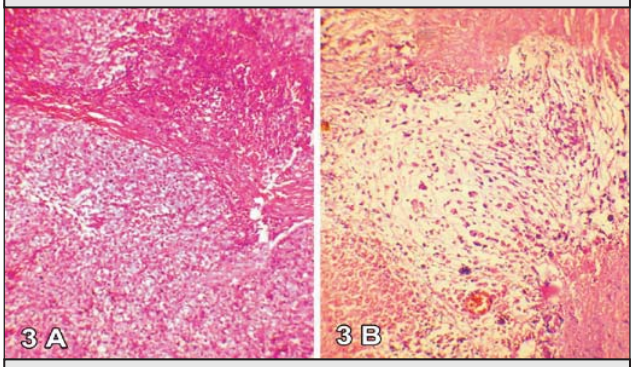

Figure 3.B. Microphotograph of recurrent lesion showing conventional chordoma with abundant amount of necrosis (H&E, X 200).

Figure 4.A. Microphotograph of FNA of left inguinal lymph node showing metastatic Chordoma cells . (Giemsa, X 200).

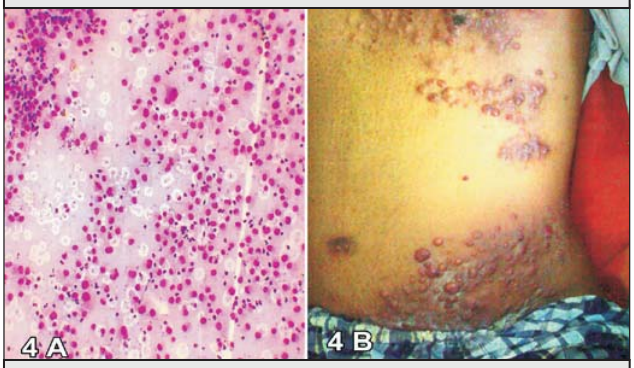

Figure 4.B. Photograph of patient showing metastatic skin lesions in chordoma involving chest wall and abdomen.

DISCUSSION

Chordomas are rare malignant tumours occurring in the midline of axial skeleton. Majority occurring in the ages of 40- 70 years (average 55) with M: F – 2:1. They are most often found arising from sacrococcygeal area (50%), skull base (35%) and the mobile spines (15%). [1, 2]

The diagnostic criteria of chordomas on fine needle aspiration include the presence of physaliphorous cells in a abundant myxoid background. [4] In this present case, fine needle aspiration of coccygeal mass showed characteristic physaliphorous cells which were helpful in establishing a preoperative diagnosis of chordoma.

Histopathologically, conventional chordomas are lobulated neoplasm, composed of polyhedral tumour cells (physaliphorous cells) in a myxoid matrix, frequently arranged in cords. [2, 6] Immunohistochemically, conventional chordoma cells typically expresses cytokeratin (CK), epithelial membrane antigen(EMA), S-100 protein and Vimentin . [10] Histopahtological analysis is required for the definitive diagnosis of chordoma. Other tumours to be considered in the Histopahtological differential diagnosis include chondrosarcoma, chondroma , metastatic mucinous Adenocarcinoma and myxopapillary ependymoma.

In our present case, the ImmunoHistoChemical findings helped in differentiating chordoma from chondrosarcoma (CK and EMA negative), metastatic carcinoma(S-100 negative) and myxopapillary ependymoma (CK and EMA negative) and supported the

theory that chordomas with its epithelial features are derivatives of the notochord.[6, 10]

Chordomas are generally thought to be locally invasive tumours of low metastatic potential.[5] Recent review of literature suggest that metastatic spread from chordoma is not an uncommon occurrence, but there is a real risk of metastatic development notable, in the case of sacrococcygeal chordoma with its incidence between 10%-48%.[1] Predicting the overall metastatic potential of chordomas generally difficult as age, gender, previous surgery and survival length have no foreseeable association. However histological anaplasia has been correlated with metastatic dissemination.[6]

Chordomas have been extensively reviewed by number of authors. They all stressed the relative rarity of tumour, its frequent sacrococcygeal location, slow, locally infiltrative and rare metastasis. Chambers and Schwinn[2] found that pulmonary (58%) and lymph node metastases (33%) are more frequent than liver(22%) and bone metastases (17%). Yarom Rana and Horn Yoan[5] found that 10% of cases of sacrococcygeal chordomas metastasize to regional lymph nodes (pre, para aortic and inguinal), lungs and liver.

Chambers and Schwinn[2] also found that sacrococcygeal chordomas have a likelihood of cutaneous involvement with incidence between 6.3% -10%. Chordomas involve the skin (chordoma cutis) by direct extension or distant metastasis. At times, metastasis to the skin may be the first sign of underlying primary tumour.[6] Clinically skin metastatic lesions of chordoma have been reported to be blue grey erythematous in colour with dermal to subcutaneous involvement .[6] In our present case ,patient developed local recurrence and left inguinal lymph node metastasis after 15 months of initial primary tumour and subsequently developed skin metastasis.

The treatment for primary tumour is surgical excision and post operative radiation therapy. However, there is a high incidence of recurrences (44% -78%) due to an inability to achieve either adequate surgical / radiation margin.[1, 6] Complete excision is rarely achieved and the usual course is one of the repeated recurrences and actually metastasis.[6] The five year survival for metastatic disease is approximately 50%.[1] Early detection is beneficial as patient outcome can improve when underlying lesion particularly in the sacrococcygeal region are removed with wide resection.

General awareness of chordomas metastasis to skin and lymph node may assist further in its recognition but by the time metastasis occurs, the typical outcome usually is poor. In describing this case we hope to emphasise the important features of chordoma that would help, facilitate the identification of this tumour and to differentiate it from other coccygeal masses and also to emphasise the rarity of lymph node and skin metastasis.

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