Case report

Aggressive osteoblastoma of the acetabulum: A diagnostic dilemma

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ABSTRACT

Osteoblastoma is a rare tumor and represents about 0.8% of all bone tumors. The aggressive variant is even rarer and difficult to diagnose. We present a case of a 17 year old male with a tumor of the acetabulum with characteristic epithelioid osteoblasts and other features of an aggressive osteoblastoma. It is important to recognize this entity as it can be clinically confused with an osteosarcoma. Our case not only showed epithelioid morphology but also had vacuolated cytoplasm, which simulated a mucin secreting adenocarcinoma. Since treatment and prognosis for these entities is strikingly different, an accurate diagnosis becomes all the more important. This case emphasizes the need for better coordination between the clinician and pathologist and use of histochemical stains and immunohistochemistry apart from routine histopathology for arriving at a final diagnosis. It is essential to have a close follow-up of the patient to look for recurrence of the tumor.

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1. Introduction

Osteoblastoma is a rare tumor and represents about 0.8% of all bone tumors.¹ Commonly affected sites are spine, diaphyses of long bones, pelvis, facial bones and short bones of the hands and feet.² Eighty per cent of the affected patients are below 30 years.¹ Despite its benign nature, the tumor may sometimes exhibit aggressive behavior.³ Dorfman defined aggressive osteoblastoma as a rare tumor that represents a borderline lesion between benign osteoblastoma and osteosarcoma. These tumors have a propensity to recur. Microscopically, they are characterized by the presence of so-called epithelioid osteoblasts and osteoid in non-trabecular pattern.⁴

2. Case report

A 17 year old male patient presented to orthopedics OPD of our hospital with complaints of pain in left hip radiating to left lower limb and difficulty in walking for the past 8 months. Patient had history of weight loss and loss of appetite. On examination, patient

had antalgic gait. The range of motion was restricted: 10–12° for flexion, 0–10° for extension, 0–30°/0–20° for abduction/adduction for the left lower limb. Distal neurovascular deficit was not seen.

X-ray showed an ill-defined non-homogenous opacity arising from medial aspect of left acetabular region and left superior pubic ramus. (Fig. 1)

CT scan revealed an expansile lytic lesion measuring 64 mm x 53 mm x 42 mm with bony septations of varying thickness in the acetabulum, superior pubic ramus and inferior aspect of left iliac blade. It had a narrow zone of transition with cortical thinning. There was evidence of irregular periosteal reaction in left iliac bone and superior pubic ramus. Femoral head appeared normal. (Fig. 2)

MR imaging revealed subtle cortical erosion of left acetabulum with altered marrow signal activity involving left acetabulum, adjacent part of left iliac blade, left superior pubic ramus and left ischium appearing hyperintense on T2 and hypointense on T1. Medial cortical outline of acetabulum was poorly visualized due to erosion with adjacent soft tissue component of size measuring 60 mm x 31 mm. Mild left hip joint effusion was seen. (Fig. 3) On MR imaging, differential diagnosis considered were

1. Infective etiology
2. Ewing's sarcoma

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Fig. 1. caption Pre-operative X-ray.
An ill-defined non-homogenous opacity arising from medial aspect of left acetabular region and left superior pubic ramus.

Patient underwent open biopsy using an ilio-inguinal approach and tissue was sent for histopathological examination. Microscopic examination revealed a tumor arranged in the form of sheets of cells in the inter-trabecular spaces. Cells were oval to plasmacytoid in shape with moderate amount of amphophilic vacuolated cytoplasm with round to oval central to eccentric moderately

pleomorphic nucleus with opened up chromatin and prominent nucleolus. Some cells showed binucleation. A vascular spindle cell stroma with abundant irregular spicules of mineralized bone and osteoid was also seen along with multinucleate giant cells. (Fig. 4)

Based on the above findings, the differential diagnosis considered on histopathology were

1. Osteoblastoma
2. Mucin secreting adenocarcinoma
3. Plasmacytoma and
4. Melanoma

Mucicarmin stain was negative ruling out an adenocarcinoma. Immunohistochemistry done was negative for cytokeratin ruling out an adenocarcinoma. Tumor cells were negative for human melanoma black 45 (HMB-45) ruling out an amelanotic melanoma. Tumor cells were negative for kappa and lambda on immunohistochemistry ruling out a plasmacytoma. However, they were positive for vimentin. (Fig. 5) Based on these findings, a diagnosis of aggressive osteoblastoma (epithelioid osteoblastoma) was offered.

In view of the diagnosis, an extended curettage was done using the same incision and excising the scar tissue. After exposure through the skin and subcutaneous tissue, abdominal muscles were split. Then the tumor was exposed using a bony window in the anterior half of the lesion. The medial wall of acetabulum was found to be intact. The contents of the lesion were evacuated and curettage of the walls was done. High speed burr was used to break septations and supplement the curette in order to remove the lesion. 80% phenol was used as an adjuvant as a pack and was left for 5 min. Adjacent obturator nerve and remaining soft tissues were protected from phenol. After phenol, ethanol was used to

Fig. 2. caption Pre-operative CT.
Shows an expansile lytic lesion measuring 64 mm x 53 mm x 42 mm with bony septations of varying thickness in the acetabulum, superior pubic ramus and inferior aspect of left iliac blade with a narrow zone of transition with cortical thinning and irregular periosteal reaction in left iliac bone and superior pubic ramus. Femoral head appears normal.

Fig. 3. caption: Pre-operative MR.

Subtle cortical erosion of left acetabulum with altered marrow signal activity. Adjacent part of left iliac blade, left superior pubic ramus and left ischium appear hyperintense on T2

Medial cortical outline of acetabulum poorly visualized due to erosion with adjacent soft tissue component of size measuring 60 mm x 31 mm. Mild left hip joint effusion seen

wash it off, followed by saline lavage. The wound was closed in layers and tissue sent for histopathological examination.

On gross examination, we received a skin covered soft tissue piece measuring 10.5 cm x 3.5 cm x 2.5 cm along with curettage of lesion. Microscopic features were similar to the previous biopsy. The skin and soft tissue were free of the tumor. One year after

extended curettage there was no clinico-radiological evidence of tumor recurrence. (Figs. 6 and 7)

3. Discussion

Aggressive osteoblastoma is a rare tumor of the acetabulum. It is characterized by:¹

Fig. 4. caption: microscopy.

4a- tumor arranged in the form of sheets of cells in the inter-trabecular spaces (H&E, magnification $\times 40$)

4b- tumor arranged in the form of sheets of cells in the inter-trabecular spaces in a spindle cell stroma (H&E, magnification $\times 100$)

4c- higher magnification showing tumor along with multinucleate giant cells (H&E, magnification $\times 400$)

4d- Oval to plasmacytoid cells with moderate amount of amphophilic vacuolated cytoplasm. Also seen is vascular spindle cell stroma. (H&E, magnification $\times 400$)

H&E: hematoxylin and eosin

Fig. 5. caption: microscopy.

- 5a- immunohistochemistry: cells positive for vimentin (magnification $\times 400$)
 5b- immunohistochemistry: cells negative for cytokeratin (magnification $\times 200$)
 5c- immunohistochemistry: cells negative for HMB-45 (magnification $\times 200$)
 5d- immunohistochemistry: cells negative for kappa (magnification $\times 400$)

1. Age range from 7 to 80 years
2. Wide and irregular trabeculae
3. Osteoid laid down in non-trabecular pattern
4. Epithelioid osteoblasts
5. Absence of cartilage

6. Osteoclast-like giant cells
7. Variable stroma
8. Low mitotic activity
9. Absence of metastases

Our case was a 17 year old male. On microscopy, we saw irregular trabeculae, osteoid in non-trabecular pattern, epithelioid osteoblasts, osteoclast like giant cells and vascular stroma. There was absence of mitosis and cartilage. No evidence of metastasis was seen on radiological imaging. Radiographs are the most helpful single imaging technique for diagnosing osteoblastoma. It may reveal varying degree of calcification.³ Our case had

Fig. 6. Post-operative photograph of the patient demonstrating the scar one year after the surgery.

Fig. 7. Follow-up X-ray..

A mixed lytic and sclerotic lesion involving left acetabulum and pubic bone. The lesion is bulging medially into pelvis with associated cortical thinning. No obvious adjacent soft tissue mass is noted.

calcification. Usually, no reactive zone of bone is seen.³ Our case had irregular periosteal reaction in left iliac bone and superior pubic ramus. It may have features similar to those of malignancy, such as cortical destruction and extra-osseous soft tissue expansion.³ Medial cortical outline of acetabulum was eroded in our case.

We report a case that had epithelioid osteoblasts and other features of an aggressive osteoblastoma. It is important to recognize this entity as it can be confused with an osteosarcoma. According to a study, the crucial differentiating features between an osteoblastoma and an osteosarcoma are:⁵

1. The absence of metastasis
2. Long term follow-up with complete resolution of the disease
3. Absence of atypical mitoses
4. Absence of cartilage cells
5. Sharp demarcation between the edge of osteoblastoma and the host lamellar bone.

The patient first presented to us in 2015 when the initial biopsy was done but was lost to follow-up. Later he returned in 2016 when extended curettage was done and since then has been on close follow-up for one year with no evidence of local recurrence or metastasis. Atypical mitoses were not seen in our case. Although our case had osteoid but it was present in a non-trabecular pattern and on microscopy, tumor was demarcated from the surrounding bone. Cartilage cells were not seen. These features helped us in ruling out an osteosarcoma.

In a study of osteoblastoma of the pelvis,⁶ it was found that pelvic osteoblastoma has a strong tendency to recur with recurrence rate varying from 24% (for osteoblastoma)⁷ to 50% for aggressive osteoblastoma,⁸ hence an extended curettage should be preferred as was done in our case.

Due to the epithelioid morphology with vacuolated cytoplasm, a mucin secreting adenocarcinoma was also considered as a differential diagnosis. Absence of mucin on mucicarmine stain helped us to rule out a mucin secreting adenocarcinoma. Other differential diagnoses considered were an amelanotic melanoma and plasmacytoma. Tumor cells were negative for HMB-45, kappa and lambda antibodies on immunohistochemistry but were positive for vimentin. Immunohistochemistry and special stains in addition to routine histopathology helped us to finally arrive at a diagnosis of aggressive osteoblastoma. It is essential to have a close follow up of the patient to look for recurrence of the tumor. One year after extended curettage, there was no clinico-radiological evidence of tumor recurrence in our case.

4. Conclusion

The clinico-radiological differential diagnoses considered in our case were Ewing's sarcoma and infective etiology. However, the pathological differential diagnoses included an osteoblastoma, adenocarcinoma, melanoma and plasmacytoma due to the epithelioid morphology of the tumor. There was complete mismatch between the clinical and pathological differential diagnosis. Finally, we arrived at a diagnosis of aggressive osteoblastoma by using immunohistochemistry and special stain like mucicarmine. This fact emphasizes the need for better co-ordination between the clinician and pathologist and use of histochemical stains and immunohistochemistry apart from routine histopathology for arriving at a final diagnosis. Although rare, a differential diagnosis of osteoblastoma should be considered in lesions involving the acetabulum. A close follow-up of a case of osteoblastoma is essential.

Sources of support

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Conflict of interest

None.

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