Unusual presentation of more common disease/injury

Ewing's sarcoma of scapula: a rare entity

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Summary

Ewing's sarcoma is the second most common malignant bone tumour of childhood and adolescence. It may affect any bone, but it is frequent in the femur, ilium and the tibia. Here the authors are reporting a case of Ewing's sarcoma involving scapula in a 14-year-old boy presenting with pain and swelling around the shoulder. Swelling was removed which on histopathological examination showed solid sheets closely packed, poorly differentiated small cells that have a high nuclear-cytoplasmic ratio, fine chromatin and small nucleoli. A tentative diagnosis of Ewing's sarcoma was made which was confirmed by immunohistochemistry.

BACKGROUND

Ewing's sarcoma of scapula is rare.

CASE PRESENTATION

A 14-year-old boy presented with pain and swelling around the left shoulder for 15 days. Initially pain was mild, persistent with no postural variations and not relieved by analgesic. The pain was associated with gradually developing swelling around the left scapular region. On clinical examination, the swelling was diffuse with ill defined margins, firm to hard in consistency and overlying skin was tense and shiny. Movements at shoulder were painfully restricted. Plain radiographs of left shoulder showed sclerotic expansile lesion of scapula with dense soft tissue shadow around the left shoulder joint (figure 1A,B). MRI of left shoulder joint was highly suggestive of Ewing sarcoma of left scapula with infiltration into the adjacent muscle mass (figure 1C,D). Surgery was done and tumour was removed which on gross examination revealed irregular brown tissue measuring about 1×0.7×0.7 cm. Cut surface was yellowish to light brown and soft in consistency. Microscopic examination showed uniform small round cell having nuclei twice the size of lymphocyte. The tumour cells had uniform round nuclei, scant cytoplasm and indistinct cellular borders. The nuclei contained finely dispersed chromatin (figure 2). Mitotic figures were also seen. No area of rosettes formation was noticed. A diagnosis of small round cell tumour favouring Ewing's sarcoma was rendered. Periodic acid-Schiff (PAS) stains showed abundant cytoplasmic glycogen (figure 3) and on the immunohistochemical stains, the tumour cell showed diffuse strong cytoplasmic membrane positivity for CD99 (figure 4). On the basis of histopathological and immunohistochemical findings a final diagnosis of Ewing's sarcoma was made. Patient was subjected to chemotherapy in the postoperated period including vincristine cyclophosphamide and actinomycin D. Patient progressed well.

OUTCOME AND FOLLOW-UP

Patient is well till now.

DISCUSSION

Ewing's Sarcoma is a primary osseous neoplasm composed of small, round, relatively uniform cells with no microscopic evidence of matrix production.¹ It comprises 6–8% of all malignant bone tumour in mayo clinic files.² The majority of patients are in the second decade of life. It shows a predilection for males.³ It may affect any bone, but is frequent in femur, ilium and tibia. The skull, vertebra, scapula and short tubular bones of hand and feet are rarely involved.⁴ Primary bone tumours and tumour like lesions of scapula are uncommon. Primary bone tumours of scapula are more likely to be malignant than benign. To the best of our knowledge less than 15 cases of Ewing's sarcoma of scapula have been reported in the literature.^{5 6} It is a malignant neoplasm constituted by a uniform proliferation of small round cells without extracellular matrix, partial or overt neuroectodermal differentiation.⁷ Ewing's sarcoma, although the second most common malignant bone tumour of children and young adults, occur rarely in scapula. The most common clinical symptoms are pain and a mass in the involved area. Fever, anaemia, leukocytosis and increase in erythrocyte sedimentation rate are often seen.⁸

Radiographically, Ewing's sarcoma of long tubular bones show an ill defined osteolytic lesion, permeative or moth-eaten bone destruction often associated with onion – slit like multilayered periosteal reaction is characteristic. Ewing's sarcoma of scapula showed considerable destructive mottling of the body of scapula, typical of a lesion with a permeative pattern with no significant bony sclerosis.

Grossly, Ewing's sarcoma is a tan-gray and often necrotic and haemorrhagic. Necrotic yellowish and semi-fluid tissue obtained from intramedullary or subperiosteal lesion might grossly be erroneously interpreted as pus by surgeons.⁹ Microscopically most cases are composed of

Figure 1 (A, B) Frontal and lateral radiograph of shoulder demonstrates diffusely sclerotic expansile lesion of scapula. (C, D) Sagittal T1-weighted MR image of shoulder shows lesion originating from scapula along with large heterogeneous extrathoracic soft-tissue mass which revealed mixed intensity signals and fluid level.

Figure 2 Small round cells having uniform, round nuclei with scant cytoplasm (H&E $\times 500$).

uniform small round cell with round nuclei containing fine chromatin, scant cytoplasm.⁸ The cytoplasm of tumour cells frequently contain PAS positive glycogen. CD99 is a cell surface glycoprotein, the product of the MIC-2 genes. CD99 is widely expressed in different tissue at a low level

but diagnostically useful in Ewing's sarcoma which typically shows a membrane positivity for CD99.¹⁰

The histologic differential diagnosis for includes small cell osteosarcoma, mesenchymal chondrosarcoma, lymphoma, metastatic neuroblastoma and primitive neuroectodermal

Figure 3 Periodic acid-Schiff stain shows abundant cytoplasmic glycogen (PAS $\times 500$).

Figure 4 Immunohistochemical stain for CD99 shows diffuse strong positivity in cytoplasmic membrane of tumour cells $\times 500$.

tumour (PNET). All the lesion share some of the histological features of Ewing's sarcoma. Morphological and immunohistochemical studies help in making the diagnosis. Distinction of Ewing's sarcoma from small cell osteosarcoma and mesenchymal chondrosarcoma is based on absence of malignant osteoid and cartilage in Ewing's

sarcoma. Lymphoma have a mixture of small and large lymphocytes with prominent nuclear convolutions and clefts, unlike the smooth contour of Ewing's sarcoma cells. Metastatic neuroblastoma generally shows rosette formation and the presence of neurofibrils. The differentiation between Ewing's sarcoma and PNET of bone is imprecise,

since the histogenous and the morphological characteristic of these tumours overlap some what.¹¹ In our patient, the tumours cytological characteristic and the absence of Homer-Wright rosettes and cytoplasmic glycogen all support the diagnosis of Ewing's sarcoma.

The general philosophy in the management of Ewing's sarcoma is to give intense multiagent neoadjuvant chemotherapy followed by enblock excision of the tumour mass. Postoperative radiotherapy is given if there is doubt of tumour residua. Important prognostic factors include the stage, anatomic location, the size of tumour, chemotherapy-induced necrosis and treatment.¹²

Learning points

- ▶ Ewing's sarcoma is a common malignant neoplasm but Ewing's sarcoma of scapula is rare.
- ▶ Clinical picture often mimic osteomyelitis.
- ▶ Final diagnosis of small round cell tumour is made after immunohistochemistry.

Competing interests None.

Patient consent Obtained.

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