Giant Ewing's Sarcoma of the Body of the Scapula

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Learning Point of the Article:
Ewing’s sarcoma of scapula is usually very aggressive in nature therefore early diagnosis and proper management is imperative for good functional outcome.

Introduction:
Ewing sarcoma is a malignant neoplasm occurring usually in bones such as femur, tibia, and iliac wing. We are here reporting a case of same arising from scapula. Of all the tumors arising from scapula, Ewing’s sarcoma contributed to only 8%.

Case Report:
A male patient presented to us with pain and swelling over right scapular region. After initial investigation, biopsy was performed. Biopsy reveals uniform small-sized cells arranged in layers with high nucleus to cytoplasmic ratio. Provisional diagnosis of Ewing sarcoma was made which was later confirmed on immunohistochemistry. The patient underwent planned surgery after neoadjuvant chemotherapy, and swelling along with partial scapula was resected.

Conclusion:
Although Ewing sarcoma is common neoplasm, it rarely arises from flat bone-like scapula. Henceforth, reporting of such a case is not only imperative but also mandatory.

Keywords:
Ewing sarcoma, scapula, malignant neoplasm, subtotal scapulectomy.

Abstract

Case Report:
A 27-year-old male presented to us with pain and swelling over right scapular region for 1 year. On local examination, a solitary huge swelling was present over the right upper back almost covering the entire scapular region, with a healed scar of previous incision biopsy done elsewhere (Fig. 1). With firm to hard in consistency, swelling was attached to underlying soft tissue and bone. Shoulder joint movements were nearly normal and there was no distal neurovascular deficit. Radiograph of the region reveals permeative pattern of destruction of scapula with no evidence of sclerosis. Routine blood workup was done. Tumor profile imaging was performed, MRI showed a large lobulated heterogeneous expansile lytic lesion indicating primary malignant bone tumor of the scapula involving scapular body with stretching of surrounding soft tissue and enlargement of axillary lymph nodes. CT scan depicted destruction of scapula bone without disturbing the glenoid cavity (Fig. 2). PET scanning was performed for metastatic
workup and showed no distant metastases. After primary imaging, core needle biopsy was performed from the lesion and histopathological examination revealed clusters of small blue round cells stacked in multiple layers with rosette and pseudorosette arrangement. Diagnosis of Ewing’s sarcoma was made after immunohistochemistry marker, CD99 and vimentin were positive. After collaborating with medical oncologist, five rounds of neoadjuvant chemotherapy were administered to the patient to counter micrometastases and to downsize the tumor. The patient underwent planned surgery. Operation was performed in floppy lateral position, posterior longitudinal incision was placed over the mass incorporating previous biopsy scar. Meticulous soft-tissue dissection was done and tumor mass was separated from all the nearby musculature with wide margins and Malawer type2 scapula resection [6] was performed, leaving behind the glenoid, acromion, and coracoid part of scapula (Fig. 3). Excised tissue was sent for examination which reciprocates the aforementioned findings of core needle biopsy (Fig. 4). Postoperatively, the patient’s arm was supported with a sling. Passive movements were started after the 2nd post-operative day. Active range of movement exercise was initiated after the end of the 3rd week. The patient was regularly followed up in oncology and orthopedic department after discharge and till now results are uneventful.

Discussion

Ewing sarcoma of bone stands second to osteosarcoma as primary bone neoplasm affecting childhood and adolescence [7]. Usually, it targets the long bones such as femur and tibia. These are Ewing sarcoma family tumors with characteristic small round blue cells on histology and “onion skin” pattern of periosteal reaction and permissive type bone destruction on radiograph [8]. Pain and swelling of the affected region comprises common symptomatology. Neoplasms originating from scapula bone are very rare. Only a few cases have been reported about Ewing sarcoma harboring the scapula. The concerned case of Ewing sarcoma occurred in late twenties of the patient with similar tissue and radiographic presentation but needs documentation as it targeted the scapula.

Battery of investigations is required to reach the diagnosis. MRI and CT scan of the region are very essential to delineate the soft tissue and bony framework, respectively, for tumor extension [9]. PET-CT has become very helpful for the assessment of distant metastases. On MRI and CT scan, in our case, tumor appears to have originated from scapular spine and has destroyed all of scapula, sparing only the glenoid framework.

In this era, with the chemotherapy and radiotherapy facilities at our disposal, limb safeguarding has become a pragmatic approach for tumors involving extremities [10]. Our patient received neoadjuvant chemotherapy to downsize the tumor and was planned for partial scapulectomy (Malawer type2). Although reconstruction with a scapular prosthesis after resection of scapula sounds more promising, we only performed subtotal scapulectomy leaving behind acromion, coracoid, and glenoid as functional outcome is better with partial scapulectomy in literatures [6].

Conclusion

“Ewing’s sarcoma of the scapula” readily pricks up surgeon’s ear, due to the rareness of the entity. Meticulous workup and a planned surgery are mandatory for its management. Further studies and case reports are required to carve out standard approaches and treatment protocols of such cases, subsequently increasing survival rate and functional outcomes for the patient.
Clinical Message

Ewing’s sarcoma of scapula is rare presentation. Thorough planning is needed for the appropriate management. Due to availability of chemotherapy and radiotherapy at our disposal, resection of such large tumors, at such surgically challenging location, is becoming possible nowadays with decreased recurrence rate and better functional outcome.

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