CASE REPORTS

SOLITARY DIAPHRAGMATIC METASTASIS OF A PREVIOUSLY EXCISED CHEST WALL CHONDROSARCOMA

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Abstract

Introduction: *Chest wall chondrosarcomas are relatively infrequent tumours. Diaphragmatic metastasis are rarer still, and are generally associated with disseminated disease, usually involving the liver and the lungs.*

The authors present the case of a 65 year old man with a prior history of a chest wall chondrosarcoma, which on a routine chest CT was identified a single diaphragmatic metastasis. A redo thoracotomy was performed, with partial resection and reconstruction of the diaphragm. The post-resection histopathological examination revealed a conventional chondrosarcoma metastasis, centred in the diaphragmatic muscle, not reaching the pleura or the peritoneum.

Keywords: *Metastasis, Sarcoma, Diaphragm*

CASE REPORT

A 65 year old man, with a prior history of a right chest wall mass involving the 3rd, 4th, 5th and 6th ribs, removed en bloc and reconstructed with methyl methacrylate "sandwich" with prolene mesh. Histological analysis showed a 25 cm, grade III conventional chondrosarcoma, with mitotic figures, some of them atypical and necrosis in over 50% of tumour area, with a minimum margin of 0.1 cm.The patient remained under surveillance.

Chest CT at 3 years revealed a de novo mass, with 50 x 30 mm involving the anterior border of the 8th rib and diaphragm, suggesting tumour recurrence. (Image 1 and image 2)

A muscle sparing anterolateral thoracotomy below the inferior border of the methyl methacrylate "sandwich" was performed. Some mild pleuropulmonary adhesions were released, the diaphragm was exposed and the mass identified on the anterior border near the cardiophrenic angle. As it was limited to the diaphragm and did not invade any other structure or organ, the diaphragmatic segment was excised en bloc (image 3) and defect was closed using a Gore-Tex graft and 2/0 prolene continuous suture. (image 4)

Histopathology showed a 55 x 45 x 30 mm conventional chondrosarcoma metastasis, with extensive myxoid areas with lymph and vascular invasion, centred in the diaphragmatic muscle, not reaching the pleura or the peritoneum.

The patient made an uneventful recovery, was discharged on the 5th postoperative day.CT follow-up at 9 months shows no tumour recurrence.

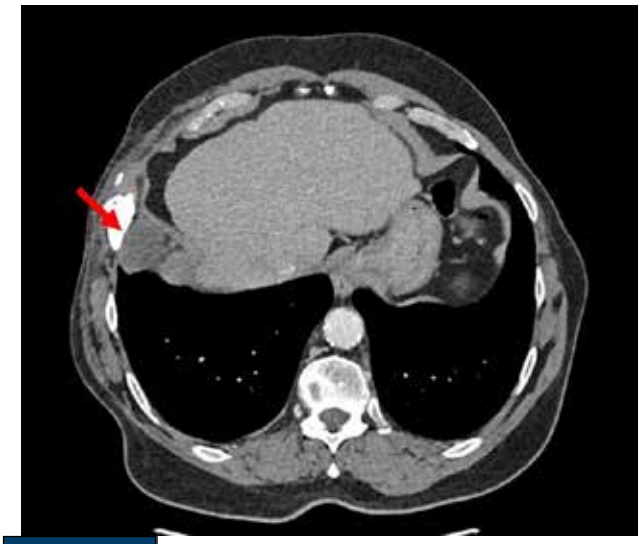

Figure 1 *Axial view CT scan showing an ovoid, homogenous, 50 x 30 mm mass, adjacent to the 8 th costal arch and diaphragm (red arrow).*

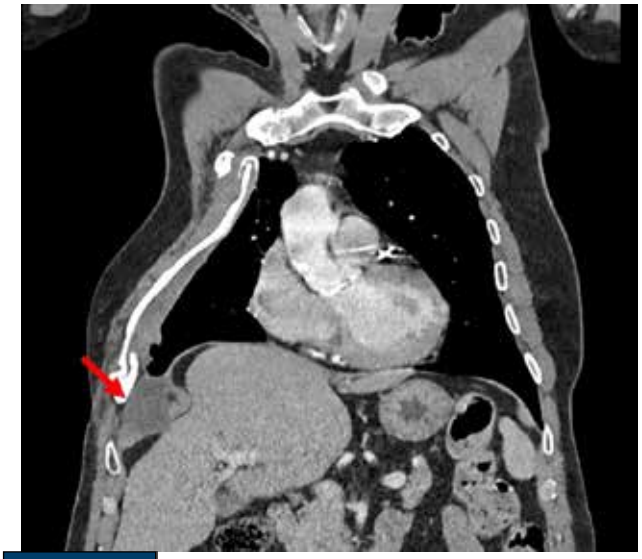

Figure 2 *Coronal view CT scan showing an ovoid, homogenous, 50 x 30 mm mass, adjacent to the 8 th costal arch and diaphragm (red arrow).*

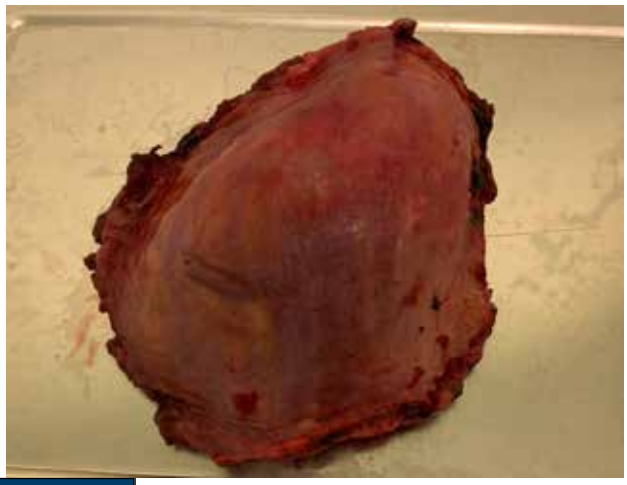

Figure 3 *Diaphragmatic segment excised en bloc containing the mass fully enclosed by diaphragm.*

DISCUSSION:

Primary central chondrosarcoma is the third most common primary malignancy of bone after myeloma and osteosarcoma. Chondrosarcoma is a tumour of adulthood and older age. The majority of patients are older than 50 years of age at diagnosis, with a slight male predominance.

The chest wall (primarily the ribs) is affected in about 15% of cases, and thus the annual incidence is extremely low for this site. However, chondrosarcoma represents the most common primary bone tumour involving the ribs and sternum[1].

Chondrosarcomas are divided into three grades based on their histopathology. Grade III tumours are pleomorphic and atypical, with peripheral cells being less differentiated and spindled, and are considered high grade. As such they tend to have a poor prognosis with rapid growth of the tumour and early metastasis, with some authors considering a rate of distant metastasis of 50 to 70 %.[2, 3]

Current NCCN guidelines recommend as first line therapy surgery whenever possible or radiotherapy. In high grade tumours with systemic recurrence or in a metastatic setting systemic therapy is recommended, but these tumours were classically thought to be resistant to chemotherapy.[4]

Recent data begun to clarify the genomic landscape of chondrosarcomas. Isocitrate dehydrogenase (IDH) mutation, found in about 50% of central chondrosarcomas, appears to be associated with higher grade and dedifferentiated tumours, portending a worse prognosis. Still these are almost completely absent in irregular bones such as vertebrae, ribs or sternum.[5]

Furthermore, positive correlation between vascular endothelial growth factor (VEGF) and VEGF-A expression and chondrosarcoma tumour grade suggests that antiangiogenic therapy may be useful in this form of bone sarcoma.[6] The SARC028 study found that anti-PD-1 antibody Pembrolizumab was able to induce a partial response in dedifferentiated chondrosarcomas, and the authors concluded that further evaluation of immune checkpoint blockade is warranted.[7]

For the time being, surgery remains the mainstay of treatment, conferring the best long term survival, but these recent advancements show great promise.

Considering the present case, diaphragmatic metastasis are rare, and generally associated with disseminated disease, usually involving the liver and the lungs. Still they are the most frequent tumours of the diaphragm, and as primary malignancies are rare.[8]

Initial impression was that of a drop metastasis from the initial surgery[9], but this was discarded as the mass was centred in the diaphragmatic muscle, adding to an extreme-

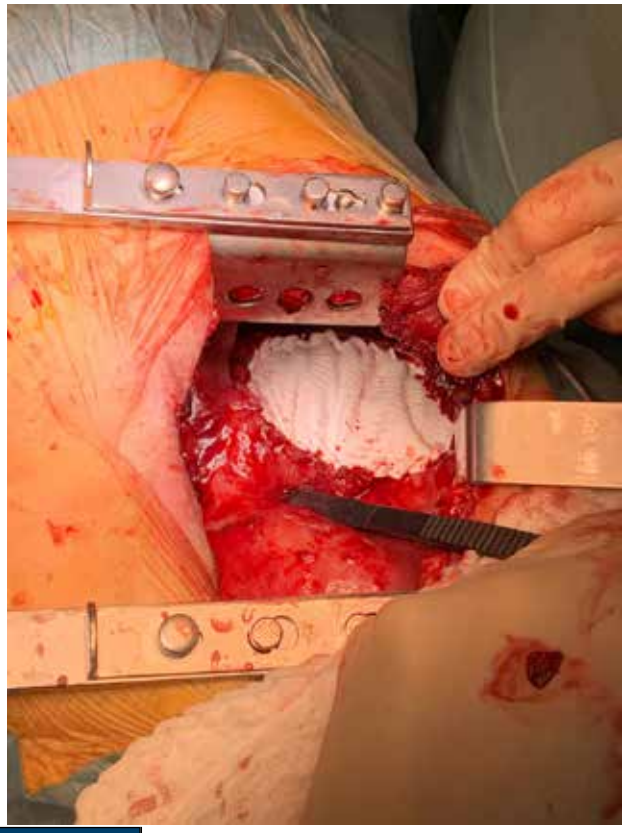

Figure 4 *Surgical image showing the reconstructed diaphragm with a Gore-Tex graft.*

ly rare occurrence, as true haematogenous spread to the diaphragm is very rare.[10]

As stated above, survival is dependent on the grade and on the location, but the majority of authors agree that a recurrence or metastasis worsens prognosis significantly.[3]

Still, it is the author's opinion that given no evidence of disease elsewhere, a single resectable metastasis of previously resected chondrosarcoma is best treated with surgical resection.

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