Metastatic synovial sarcoma of the sternum: A case report and review of the literature

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Synovial sarcomas and metastasis of sarcomas to the sternum are a rare occurrence. Due to the rarity of occurrence, a case of synovial sarcoma metastasizing to the sternum with reconstruction is reported.

Key Words: *Metastatic sarcoma; Sternum*

Sarcome synovial et métastase au sternum : rapport de cas et revue de la littérature

RÉSUMÉ : Les sarcomes synoviaux et les métastases sternales des sarcomes sont des phénomènes rares. Compte tenu de la rareté de ce tableau, nous présentons ici un cas métastase sternale d'un sarcome synovial avec reconstruction.

Synovial sarcomas are rare tumours that occur in the vicinity of joint and tendon sheaths of the extremities. Primary tumours of the sternum are very rare, and metastatic lesions have approximately the same incidence as that of primary tumours. Metastatic tumours at the sternum originate from thyroid, kidney, lung, breast or prostate tumours (1-4). A review of the literature did not reveal any report of metastatic synovial sarcoma to the sternum.

CASE PRESENTATION

A 42-year-old man presented eight years after an above-knee amputation with tenderness and swelling of the sternum following a fall. He had a left above-knee amputation for synovial sarcoma around the knee, followed by radiation in 1987. He later developed a solitary lesion in his left lung for which he underwent a left thoracotomy and lower lobe resection in 1991. Histology of this lesion confirmed that it was a metastatic synovial sarcoma. He was asymptomatic and had been followed in the oncology clinic for nearly four years before this fall. The swelling involved the body of the sternum (Figure 1). Because his chest x-ray showed a soft tissue mass, a computed tomography (CT) scan was done (Figure 2). A clinical diagnosis of metastatic disease was made. Because the lesion involved the whole

body of the sternum, no preoperative biopsy was carried out. He underwent excision of the sternum. On gross examination (Figure 3), the whole body of the sternum was replaced with the tumour. The lesion measured 11×5 cm. There was cystic degeneration, with tumour necrosis in an area measuring 2.5 cm². The lesion infiltrated into the chest wall fascia, muscle and chondral portion of the first and second ribs. Histology con-

Figure 1) *Swelling of the sternum due to the presence of a tumour*

Figure 2) *Computed tomography scan of the patient's chest wall*

Figure 4) *Histology of the lesion showing synovial sarcoma*

Figure 3) *Gross lesion of the sternum*

Figure 5) *Marlex mesh in place*

firmed the lesion to be biphasic with a mitotic index of 2.5 per 10 high power microscopic field and similar to the lesion from his left lower extremity. There were cuboidal cells surrounded by fibrosarcoma-like spindle cell elements (Figure 4). The sternal defect was reconstructed using two layers of Marlex mesh (CR Bard Inc, USA) sutured to the ribs as a tie-beam (Figure 5), and the pectoralis muscles were transposed

over the mesh. On the nondominant upper extremity, the pectoralis muscle was divided from its insertion to the humerus to cover the lower end of the defect (Figure 6). The skin incision was closed over the muscle (Figure 7). The patient underwent postoperative radiation to his chest wall. The patient was disease free 24 months after surgery with a stable chest wall.

Figure 6) *Transposed pectoralis muscle flaps*

DISCUSSION

Synovial sarcoma, also called synovioma, is a rare cancer that begins in the synovial tissue. This sarcoma is a clinically and morphologically well defined entity. Although extensively described, its biological features are controversial. Although it has been found in areas remote from joints, such as the neck and abdominal wall, it is considered to occur in the vicinity of joint bursae or tendon sheaths, especially in the knee region. This cancer occurs mainly in adolescents and young adults, and affects more males than females. The most common symptom of synovial sarcoma is a deep-seated swelling or a mass that may be accompanied by pain or tenderness. The prognosis has been correlated to tumour size, histological subtype, mitotic rate and tumour necrosis (5,6). These tumours arise from the mesenchyme rather than mature synovial tissue, which has epithelial and supporting tissue features. Accordingly, their histological types can be epithelial, stromal or mixed. Two types of synovial sarcoma are commonly classified by light microscopy: the predominantly biphasic type and the predominantly monophasic type. The biphasic type, is characterized by the appearance of spindle-cell stroma and gland-like slits lined by what appear to be epithelial cells. The columnar or cuboidal epithelial cells are surrounded by fibrosarcoma-like spindle cell ele-

Figure 7) *Wound closure with mediastinal drains*

ments. The monophasic type may be categorized as 'low glandular' or 'high glandular', depending on the spindle cell pattern. Although there is no agreement as to the prognostic significance of the histological subtype, the biphasic biological pattern has been associated with a better survival rate (1,5). Synovial sarcoma is known to metastasize late (7), and the biological activity is independent of the anatomic site of origin. The incidence of distant metastasis was 49%, and tumour size was a highly significant prognostic feature (1).

Metastatic disease is diagnosed by CT scan and biopsy. Bone marrow scintigraphy has been reported to be useful in the early detection of tumour invasion into the bone marrow at an early stage before the destruction of skeletal tissue. The removal of the primary tumour and repeated pulmonary recurrence are rewarding to the patient because the patient can live a disease free life until another lesion arises (8). The aim of surgery for a sternal tumour is to remove the growth with a reasonable excision margin. Extensive resections of sternum and peristernal structures obviously alter respiratory dynamics. Five year survival rates are misleadingly optimistic and do not adequately reflect the natural history of the sarcoma. If local excision margins are insufficient, excision should be performed, followed by adjuvant radiotherapy and chemotherapy.

Synovial sarcomas tend to recur locally, and distant metastasis may occur many years after the initial diagnosis. Because these tumours can metastasize many years following resection, these patients should be carefully followed up.

Since the first report of partial sternectomy by Holden (9) in 1878, most of the early reports advise preservation of part of the sternum for chest wall stability. Extensive resections of sternum and chest walls are feasible provided that chest wall repair affords enough stability and airtight closure. To obtain chest wall stability, reconstruction of the sternum should be carried out following a radical resection. This procedure may be done using autologous rib grafts or prosthetic materials.

Common prosthetic materials used are prolene mesh, Marlex mesh, and methylmethacrylate sandwiched between two layers of prolene (10). While chest wall stability may be achieved with prosthetic materials, the soft tissue coverage may be obtained using local muscle flaps, myocutaneous flaps or omentum. In this patient, reconstruction and airtight closure was obtained using Marlex mesh and bilateral pectoralis muscle flaps.

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