CASE REPORT

Extraskkeletal myxoid chondrosarcoma of the neck

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Extraskkeletal myxoid chondrosarcoma is a rare malignant soft-tissue tumour that is typically in the deep soft tissues of the lower extremity. The tumour is usually a well-defined, multinodular soft-tissue mass without calcifications. A 62-year-old woman with a history of nasopharyngeal cancer presented with a palpable mass in the anterior neck. Radiologically, the lesion was a well-defined soft-tissue mass with the extensive calcifications on various imaging examinations. Although this lesion was histopathologically diagnosed as extraskkeletal myxoid chondrosarcoma, the unusual imaging findings were challenging and very intriguing.

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Case report

A 62-year-old woman presented with a palpable mass in the right anterior neck which had gradually enlarged during a period of 3 months. 9 years earlier, she had had an undifferentiated nasopharyngeal carcinoma (T1N2M0), which was treated with a combination of concomitant chemotherapy and radiotherapy. The total dose of radiation was 6660 cGy given in 40 fractions. No recurrence was found at follow-ups.

A CT scan showed a soft-tissue mass in the right internal jugular chain that measured 5.0 × 3.0 cm in diameter. The mass was well demarcated from the surrounding structures and had dense calcifications (Figure 1).

Ultrasonography revealed a well-defined, solid echogenic mass containing multiple calcifications with posterior acoustic shadowing (Figure 2).

MRI showed that the mass was of low signal intensity on a T_1 weighted image (Figure 3a). On a T_2 weighted image, the peripheral area of the lesion showed high signal intensity, while the central portion showed low signal intensity (Figure 3b). Following contrast injection,

the peripheral portion corresponding to the high signal intensity on T_2 weighted images revealed strong enhancement, except the central portion (Figure 3c).

An ^{18}F -fluorodeoxyglucose whole-body positron emission tomography and CT (^{18}F -FDG PET/CT) scan demonstrated an ill-defined peripheral hypermetabolic rim of the lesion (Figure 4).

Initial differential diagnosis before surgery was thyroid cancer and metastatic lymphadenopathy.

The tumour was widely excised surgically. The specimen was a circumscribed solid mass measuring 6.0 × 3.5 cm and there was a yellow-white matrix on the cut surface. Microscopically, the tumour exhibited lobular structures of atypical spindle cells, embedded in an abundant chondromyxoid matrix (Figure 5). These findings were considered to be consistent with the diagnosis of extraskkeletal myxoid chondrosarcoma.

Discussion

Extraskkeletal myxoid chondrosarcoma is a rare soft-tissue sarcoma with ultrastructural and molecular features distinct from that of skeletal myxoid chondrosarcoma.^{1,2} Enzinger and Shiraki first described it in 1972 as a distinct clinicopathological entity

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Figure 1 Transverse CT scan showed the enhancing soft-tissue mass in the right side of the neck containing multiple calcifications (long arrows). The right internal jugular vein has been compressed by the mass (short arrow)

with a sarcoma composed of primitive chondroid cells in an abundant myxoid matrix resembling embryonic cartilage.^{3,4} Extraskelatal myxoid chondrosarcoma had a protracted clinical course characterized by a high rate of metastases and local recurrences and eventual tumour-related death in a high percentage of cases.¹⁻⁴ The tumour frequently arises in the deep soft tissues, especially in the lower extremities. Other sites that have been reported include the buttock, shoulder and hand.^{1,2} There are a very few reports of extraskelatal myxoid chondrosarcoma in the head and neck.⁵

Figure 2 Longitudinal ultrasound image of the right side of the neck confirmed the calcifications in the mass (arrows)

On clinical examination, extraskelatal myxoid chondrosarcoma has no specific findings. In most cases, the neoplasm occurred as a single slow-growing, soft-tissue swelling, usually measuring 5–10 cm in diameter.³ Our patient had a gradually enlarging mass in her neck, which may have existed for 3 months.

On CT scans, extraskelatal myxoid chondrosarcoma appears as a dense soft-tissue mass with lobular contour and no distinctive radiological features.^{5,6} Calcification is usually absent or if present it is not prominent.⁵⁻⁷ In our case, prominent calcifications were detected in CT scans and ultrasonography. Therefore, the extensive calcification as seen in this case is a rare imaging finding not previously reported.

Extraskelatal myxoid chondrosarcomas were best visualized on MRI, in which the presence of myxoid stroma could be readily appreciated.¹ Characteristic imaging features of these tumours are soft-tissue masses, presenting hyper-signal intensity on T_2 weighted MR images.² However, the tumour in our case showed a central low signal intensity with a peripheral high signal intensity on T_2 weighted MR images and a peripheral rim of enhancement after administration of a contrast material, which might be owing to multiple calcifications in the central portion of the mass.

Our case showed a peripheral hypermetabolic rim of the tumour with a central metabolic defect on PET/CT, which is similar to that on post-contrast MR images. To the best of our knowledge, PET/CT of extraskelatal myxoid chondrosarcoma has not previously been described in the radiological literature.

The differential diagnosis of the malignant soft-tissue tumours of the neck is broad and includes malignant fibrous histiocytoma, liposarcoma, synovial sarcoma and fibrosarcoma. Malignant fibrous histiocytoma and myxoid liposarcoma are representative myxoid soft-tissue tumours, but there were no further characteristics of a particular type of myxoid soft-tissue tumour.^{8,9} Malignant fibrous histiocytomas are often of homogeneous soft-tissue attenuation on CT but may exhibit small foci of calcification, haemorrhage or necrosis. On MRI, they have internal low-signal septations and a heterogeneous high signal intensity on T_2 weighted images.^{9,10} Sundaram *et al*⁸ reported that lacy or linear foci of high signal with a low-signal mass on T_1 weighted images may permit the presumptive MRI diagnosis of myxoid liposarcoma.⁸ When only non-lipomatous material is present, myxoid liposarcoma cannot be distinguished from other soft-tissue tumours by radiological imaging.

In conclusion, we have reported a recent case of an extraskelatal myxoid chondrosarcoma of the neck. The current report is unique in several aspects, including CT and sonographic appearance of extensive calcifications, unusual MRI and PET/CT features and a rare location in the neck region. Although these findings may be seen in other soft-tissue neck malignancies, an extraskelatal myxoid chondrosarcoma should be considered a possibility in differential diagnosis in soft-tissue neck mass.

a

b

c

Figure 3 (a) MRI revealed a well-defined mass in the right neck with slightly low signal intensity on T_1 weighted images and (b) increased signal in peripheral portion with a low-signal central portion on T_2 weighted images. (c) Strong peripheral rim-enhancement of the mass was observed on post-contrast T_1 weighted images

Figure 4 Axial positron emission tomography (PET)/CT image demonstrates a mild peripheral metabolic rim of the calcified neck mass (arrows). Intense bilateral thyroid hypermetabolism is also noted

Figure 5 Histological section of the resected specimen shows a solid mesenchymal tumour composed of diffusely proliferating spindle tumour cells and immature chondroid or cartilaginous elements in myxoid stroma (haematoxylin and eosin, $\times 100$)

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