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Undifferentiated pleomorphic sarcoma of the neck – A case report

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ABSTRACT

Undifferentiated pleomorphic sarcoma very rarely affect the neck. We report a case of a 62 year old man who presented with a right sided skin lump which he noticed after sustaining a neck contusion in a road traffic accident about one year ago. The initial CT and ultrasound scans of the lump were suggestive of a thrombosed varix. Clinical examination showed a hard skin nodule with signs of tethering. He underwent a wide excision of the skin nodule and the histology showed undifferentiated pleomorphic sarcoma with margins involved. He subsequently underwent another re-excision of margins and pectoralis major flap reconstruction. Following surgery, he was also prescribed adjuvant radiotherapy and he remains well about 12 months after follow up. The surgical management of undifferentiated pleomorphic sarcoma in the neck is challenging due to the proximity of critical neck structures and the need to obtain clear margins. Thus, adjuvant radiotherapy is often used to improve disease control.

1. Introduction

Sarcomas of the head and neck region are uncommon, with over 50 histological subtypes and represent less than 1% of all cancers in the head and neck area [1,2]. Undifferentiated pleomorphic sarcoma is a high grade aggressive tumour which is notoriously difficult to obtain surgical clearance due to its proximity to important structures in the neck.

Our work has been reported in line with the SCARE criteria [3].

2. Case report

A 62 year-old gentleman was referred by his family physician for a possible sebaceous cyst at the right neck which had further increased in size over the last 3 months. He first noticed the skin lump after sustaining a superficial injury to the right neck during a road traffic accident about a year ago. Magnetic resonance imaging then showed an ill-defined T2 hyperintensity in the lower end of the right sternocleidomastoid muscle that was mildly contrast enhancing. There was also a sliver of fluid adjacent to it. This was suggestive of a possible contusion. He did not have any prior irradiation to that neck region nor any significant medical and family history of note. He is a non-smoker.

On examination, he had a 2.5 cm smooth, hard nodule in the right upper supraclavicular region (Fig. 1). The skin superiorly was thickened and appeared tethered to the underlying nodule. There was no cervical lymphadenopathy.

CT (computed tomography) neck (Fig. 2) showed a 2 cm soft tissue nodule in the right lower neck region confined to the subcutaneous tissue. It was in close proximity to a small branch of the right anterior jugular vein and had no intramuscular extension. A likely differential was a thrombosed venous varix.

Subsequently, an ultrasound scan of the neck (Fig. 3) showed a $2 \times 1.5 \times 2.3$ cm heterogenous solid nodule attached to the skin with internal vascularity. It laid close to the external jugular vein (EJV) and splayed the EJV's tributaries but there was no gross vascular invasion or thrombosis.

The patient was scheduled for a wide excision of the nodule by a Head and Neck surgeon to exclude a skin malignancy in view of the unusual finding of skin tethering. Intraoperatively, the nodule was found to be lying in the subcutaneous tissue and over the sternocleidomastoid (SCM) muscle. The overlying skin was tethered by a cord-like extension from the nodule. The frozen section of the nodule revealed a malignant spindle cell tumour and hence additional skin and SCM margins were taken. These samples showed atypical cells and thus a decision was made to await the final histology results before deciding if further resection was needed prior to elective flap coverage in a few days' time. While awaiting the histology results, the patient was nursed in the ward with regular dressing to his neck wound. A staging CT thorax and abdomen

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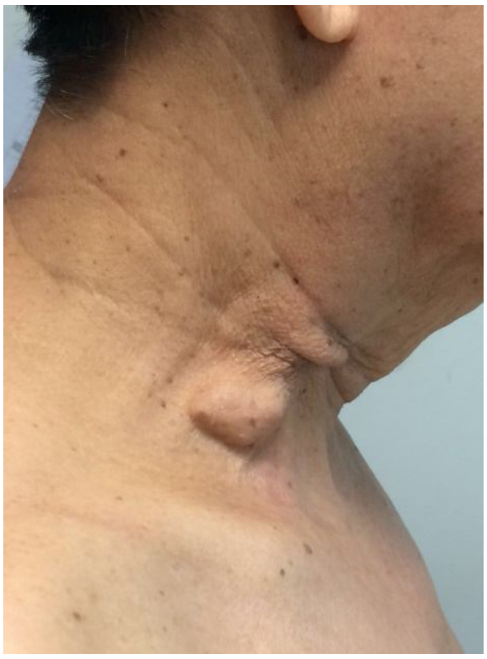

Fig. 1. Photograph of the right neck mass.

was done prior the second surgery and the scans did not reveal any distant metastases.

The final histology showed a malignant tumour located in the dermis and subcutaneous tissue. The tumour comprises predominantly spindle cells arranged in fascicles. The cells had enlarged irregular hyperchromatic nuclei and moderate eosinophilic. Brisk mitotic activity was also noted. Immunostains were negative for cytokeratins (AE1/3), CD34, h-caldesmon, desmin and HMB45. The diagnosis was that of T1b Grade 3 undifferentiated pleomorphic sarcoma with the medial and deep margins involved. The superior and lateral margins were noted to be <1 mm. Hence, the patient underwent a re-excision of skin margins by the Head and Neck surgeon again and reconstruction with a pedicled right pectoralis major myocutaneous flap by a Plastics surgeon. During the surgery, the great auricular nerve (GAN) was noted to be adjacent to where the SCM had been excised in the previous surgery. This short segment of GAN was separately excised for histology. Level II to IV nodes were taken en bloc with the right sternocleidomastoid muscle, internal jugular vein and spinal accessory nerve. The second histology results showed that all margins and lymph nodes were cleared of malignancy except for the presence of tumour cells on the GAN. The patient made an uneventful recovery after the second surgery and was discharged about one week later.

This case was discussed at a multidisciplinary tumour board and the patient subsequently opted for adjuvant radiotherapy. He tolerated the adjuvant radiotherapy well and at one year post treatment, surveillance CT scans of the neck and thorax reported no evidence of tumour recurrence.

3. Discussion

Undifferentiated pleomorphic sarcoma was previously known as malignant fibrous histiocytoma and is an uncommon sarcoma in the head and neck [4] as it usually afflicts the extremities. The commonly described presentation of head and neck sarcomas is that of a painless mass [2]. Undifferentiated pleomorphic sarcoma arises from mesenchymal cells and typically presents in patients in the 50–70 age group. Diagnosis requires tissue from a needle biopsy or occasionally, an excisional biopsy for smaller tumours. Immunohistochemistry is often required to exclude tumour arising from muscle, neural and adipose differentiation, as well as a sarcomatoid tumour. In our patient, the appearance of a neck swelling after trauma and its features on CT suggested that the nodule could be a thrombosed varix. However, what was unusual about the nodule was its slow, progressive increase in size and the tethering of the overlying skin. Thus, surgical excision for histology was recommended to the patient.

Due to the scarcity of undifferentiated pleomorphic sarcoma, most of the medical literature report head and neck sarcomas as a whole. Surgical excision with clear margins forms the mainstay of treatment for this condition. A comparison of surgical and non-surgical treatment revealed that patients who underwent surgery had greater 5-year local control (56% vs 40%) and 5-year survival rates (70% vs 40%) [5]. However, clear surgical margins can be difficult to achieve – Lindford *et* al. looked at 39 head and neck sarcoma patients who underwent surgery and found that 12.8% of the patients had close margins (<1 mm) while 23.1% had involved margins [6].

The role of radiotherapy lies mainly in improving local disease control and overall patient's survival after tumour resection. A study by Colville et al. showed that surgery with clear margins combined with adjuvant radiotherapy had significant improvement in 5-year local control, 5-year disease-free survival and 5-year survival compared with surgery alone [5]. However, this benefit was less if surgical margins were less complete [5]. Interestingly, a study by de Bree et al. found that adjuvant RT mainly increases the rate of local control but not for overall survival as patients subsequently develop distant metastasis [1]. Nonetheless, a study by Mahmoud et al. found that overall survival was improved in patients who underwent adjuvant RT for high grade sarcomas [7]. This suggests that radiotherapy should be considered in patients who have high grade sarcomas.

In a study by Vitzhum et al. which analysed their cases retrospectively, neoadjuvant radiotherapy was mainly used in patients who had marginally resectable tumours [8]. In this study, a comparison between patients who underwent neoadjuvant radiotherapy and those who underwent adjuvant radiotherapy showed no difference in overall survival. However, patients who had adjuvant radiotherapy had higher risk of fibrosis and swelling while patients who had neoadjuvant radiotherapy were found to have higher rates of severe wound complications.

Radiotherapy is also sometimes utilized as a primary treatment modality in patients who are medically unfit or have inoperable tumours [5]. Not surprisingly, Radiotherapy (RT) without surgery is associated with lower locoregional control, disease free survival (DFS) and overall survival (OS) [8].

Besides margins status, some reports suggest that grade and a tumour size more than 5 cm were prognostic factors for survival [1,9–11]. Tumour grade is more influential than tumour size as a high grade sarcoma has greater potential for distant metastasis [1].

Although PET imaging is not routinely used in the initial staging of pleomorphic sarcoma [12], the role of PET scan as a prognostic tool in soft tissue sarcoma is evolving. Being a metabolic imaging, PET can identify areas of necrosis in high grade sarcomas by virtue of their increased FDG uptake and thus predicts overall survival [13,14] as well as disease free survival in patients [14]. Lastly, PET scan may have a role in assessing tumour response to neoadjuvant treatment [15,16].

4. Conclusion

Sarcomas of the head and neck region are rare and clinical features of skin infiltration such as tethering by a skin nodule should raise one's concern of an underlying malignancy, despite

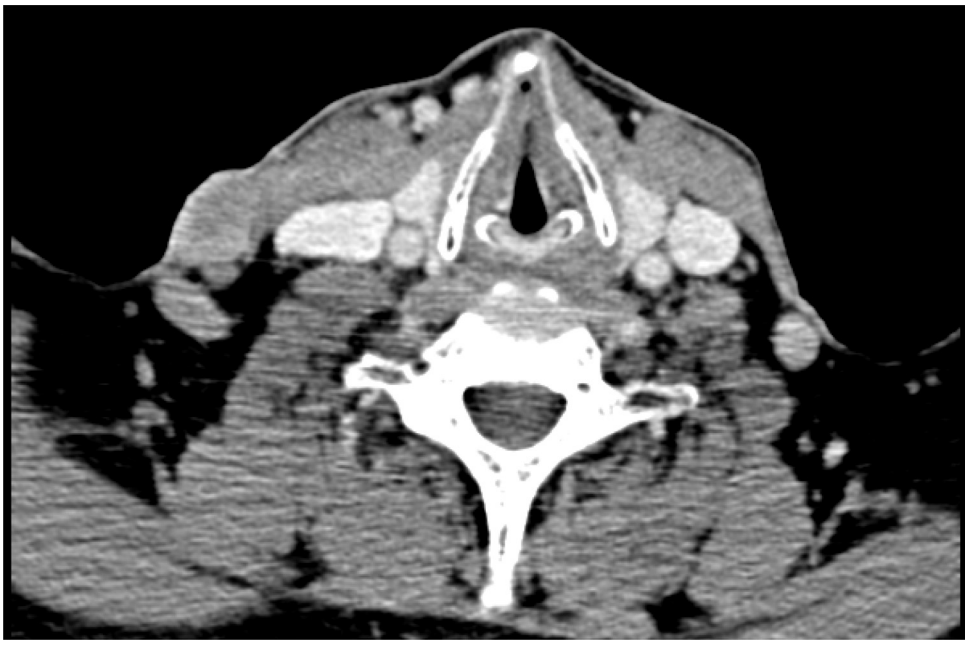

Fig. 2. CT showing the subcutaneous nodule.

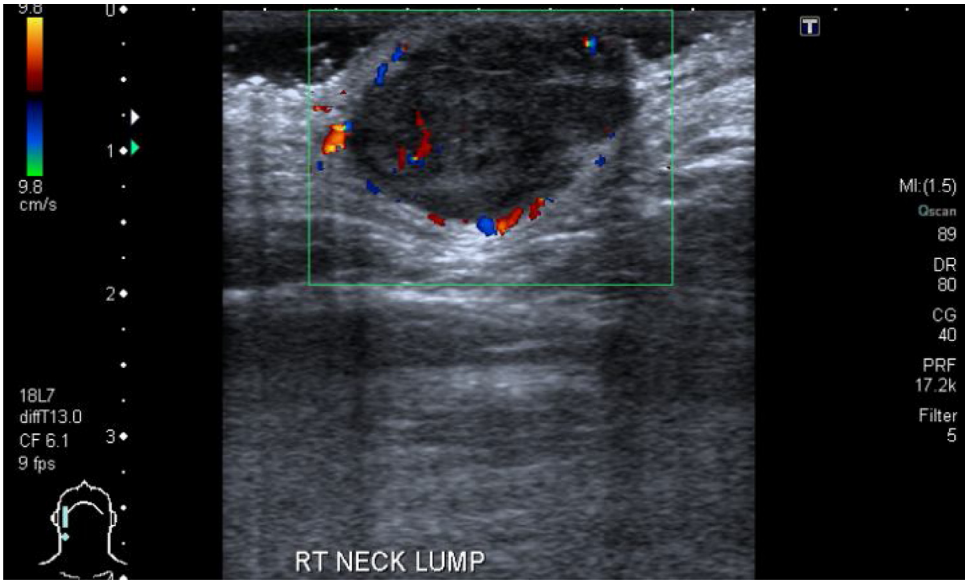

Fig. 3. US showing the same nodule with internal vascularity.

the absence of suspicious features on radiological imaging. Achieving clear margins during surgery may be difficult given the lesion's proximity to vital neck structures. Hence, a multidisciplinary approach to treatment is necessary and this entails tumour extirpation by a resection surgeon, soft tissue coverage by a reconstructive surgeon and subsequent adjuvant radiotherapy prescribed by a radiation oncologist if surgical margins are involved.

Conflicts of interest

All authors do not have any conflict of interest.

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Ethical approval

This is a case report and not a research study and therefore no ethics committee is involved.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Jane Peiwen Lim – case report write up.
Jack Kian Chng – review of images.

Choon Chieh Tan – review of case write up.

Guarantor

Guarantor- Choon Chieh Tan (Dr).

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