Supraclavicular Atypical Lipomatous Tumour: Lesson Learnt

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ABSTRACT

Sarcomas are malignant mesenchymal neoplasms that comprise less than 1% of all malignancies diagnosed annually in all body sites; a number far surpassed by epithelial-derived carcinomas. Of the soft tissue sarcomas, liposarcoma, derived from adipose tissue, accounts for approximately 17% to 30% of all soft tissue sarcomas and is considered one of the most common soft tissue sarcomas in adults. Head and neck liposarcoma are markedly less common than those in other body sites, comprising only 2% to 9% of sarcomas. We herein report the case of a 62-year-old male with a two year history of a right supraclavicular mass, which turned out to be liposarcoma and its further management. A missed step in the algorithm of head and neck swelling management that is fine needle aspiration cytology (FNAC) precipitated a less than ideal management in this case.

Keywords: Liposarcoma; Atypical Lipomatous Tumour; Head and Neck Liposarcoma

INTRODUCTION

Sarcomas are malignant mesenchymal neoplasms that comprise less than 1% of all malignancies diagnosed annually in all body sites; a number far surpassed by epithelial-derived carcinomas [1]. Of the soft tissue sarcomas, liposarcoma, derived from adipose tissue, accounts for approximately 17% to 30% of all soft tissue sarcomas and, is considered one of the most common soft tissue sarcomas in adults. Head and neck liposarcoma is markedly less common than in other body sites, comprising only 2% to 9% of sarcomas found in the region [1-2]. Its rarity is also illustrated by the fact that at a comprehensive cancer institute, only 30 patients were treated for head and neck liposarcomas during a 60-year period [3].

The presumed origin of liposarcomas from totipotential primitive mesenchymal cells would account for the wide variety of cell types that compose these tumours, including mucoid lipoblasts; signet ring cells; immature, mature and well-differentiated lipocytes; mulberry cells with a round central nucleus and many cytoplasmic vacuoles; as well as bizarre giant cells [4]. The World Health Organization (WHO) divides liposarcomas into dedifferentiated, well-differentiated (also often called atypical lipomatous tumour, due to the absence of metastatic potential), myxoid, pleomorphic and mixed-type liposarcomas [5, 7]. Well-differentiated liposarcoma is an intermediate (locally aggressive) malignant mesenchymal neoplasm composed either entirely or in part, of a mature adipocytic proliferation showing significant variation in cell size and at least focal nuclear atypia in both stromal cells and adipocytes [5].

This case report highlights a 62-year-old male with a two years history of a right supraclavicular mass, which turned out to be liposarcoma. A missed step in the algorithm of head and neck swelling management, that is fine needle aspiration cytology (FNAC), precipitated a cascade of events resulting in less than ideal management in this case.

CASE REPORT

A 62-year-old male presented to the Ear Nose Throat (ENT) clinic of the Penang General Hospital with a two-year history of a right supraclavicular mass. The mass had rapidly increased in size from about 1cm x 1cm to about 5cm x 4cm over the duration of three months

prior to presentation. He had discomfort with his right arm movement since the size increased. He had type II diabetes mellitus, hypertension and dyslipidemia. On examination, the swelling was in the right supraclavicular fossa with no palpable cervical lymphadenopathy. The swelling was not fixed to the underlying muscle, was non-tender, and had a smooth surface on palpation. Slip test was negative. The overlying skin was normal. The ENT examination of the patient was unremarkable. He had a full range of movement of his right arm and there were no motor or sensory neurological deficits.

MRI of right supraclavicular fossa revealed a mass within the right trapezius muscle, measuring 5.2cm x 3.2cm x 2.5cm in size. It was ovoid with well-defined but lobulated margins. It was isointense to muscle on T1 weighted image (Figure 1) and hyperintense on T2 weighted image. It enhanced fairly homogenously with contrast. Low signal within the mass on T2 may have indicated an area of haemorrhage or calcification within the mass. The mass was confined to the trapezius muscle belly. The radiological appearance was suggestive of a primary neoplasm such as a rhabdomyosarcoma. Fine needle aspiration cytology (FNAC) was not performed, as given the clinical presentation at that time, it was suspicious of a benign lesion and we had operating theatre time for excision within few days of patients' presentation. We decided to proceed with excision after discussion with our orthopedic surgeons.

An S-shaped incision was made over the swelling, which extended from the anterior part of trapezius muscle laterally towards the clavicle bone. The mass (Figure 2) was easily separable from the underlying structures and from the trapezius muscle and was removed as a whole. The tumour was situated inferior to the most anterior bulk of trapezius muscle, which explains the disconcordance between the MRI and clinical findings. The brachial plexus and the spinal accessory nerve were intact.

Pathological examination revealed a yellowish ovoid shaped mass measuring about 5cm x 3cm macroscopically. Microscopically, the section showed a well-circumscribed partly and thinly encapsulated lesion formed by variably sized adipocytes arranged in lobules separated by fibromyxoid septa containing atypical stromal cells. Occasional multivacuolated lipoblasts were seen. The scattered atypical stromal cells exhibiting marked nuclear hyperchromasia and enlargement with abundant eosinophilic cytoplasm were also seen. Loose myxoid areas with curvilinear vascular channels were observed but no round cells were appreciated. CD34 staining was positive in the atypical stromal cells. Epithelial membrane antigen (EMA) and smooth muscle actin (SMA) stainings were negative. The lesion was interpreted as atypical lipomatous lesion (well-differentiated liposarcoma) with involved inferior, lateral, posterior and anterior margins (Figure 3-6).

The patient received adjuvant radiotherapy as the margins were involved. He was not further subjected to local wide excision as it could lead to functional morbidity and there were vital structures surrounding the margins involved. He has completed his treatment and follows up monthly at our clinic.

DISCUSSION

Lipomatous neoplasms are commonly encountered by the general surgeons as well as the surgical oncologist. Collectively, the lipomatous neoplasms comprise half of all soft tissue tumors, but the individual histologic subtypes are various and range from the benign lipoma to the aggressive liposarcoma. Although an experienced surgeon can frequently distinguish the benign lipoma from the aggressive liposarcoma, the fine distinction between large lipomas and similarly large atypical lipomatous tumors creates a more challenging diagnostic dilemma [5]. Accurate recognition and differential diagnosis of lipomatous tumours are of major importance for adequate treatment. This is where the FNAC plays a major role. The biological behaviour of lipomatous tumours varies greatly and need to be treated accordingly. Lipomas are usually treated with simple excision. Some other benign lipomatous tumours are preferably treated by wide surgical excision because of their high local recurrence rate after simple excision. In planning for these cases, it is essential to balance the completeness of the procedure with the risk of unnecessary functional and cosmetic morbidity. On the other hand, liposarcomas require resection with wide margins for adequate treatment. In some cases, radiotherapy or chemotherapy may precede surgery [4, 8]. As our patient fell in the liposarcoma category, an early and proper planning would have allowed an excision with a clear margin.

MRI was done in this case with the suspicion of soft tissue tumour as the MRI has become technique of choice for identification and characterization of soft tissue tumours [9]. MRI

Figure 1: Coronal T1-weighted MRI of the mass within right trapezius muscle

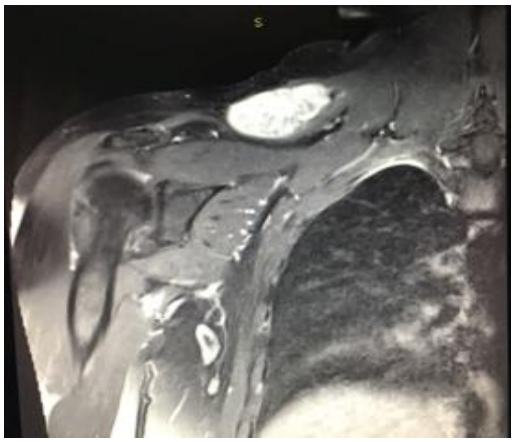


Figure 2: Right Supraclavicular mass

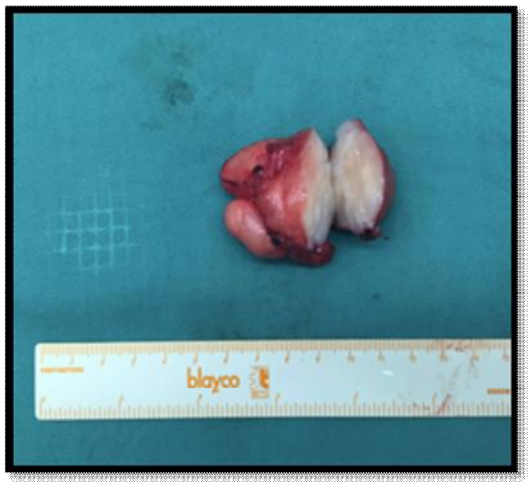


Figure 3: Tumor composed of adipocytes with myxoid stroma containing atypical stromal cells. H&E stain.
X4 magnification

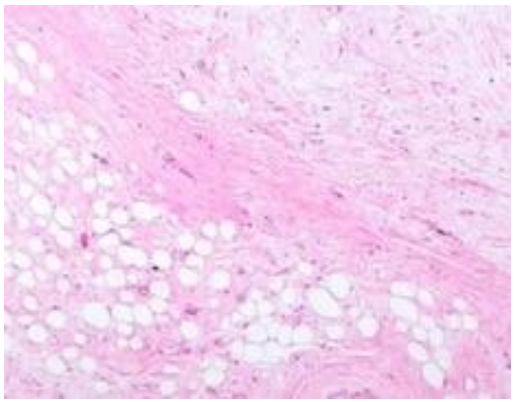


Figure 4: Multivacuolated lipoblast (red arrow) and univacuolated lipoblast (brown arrow). H&E stain.
X40 magnification

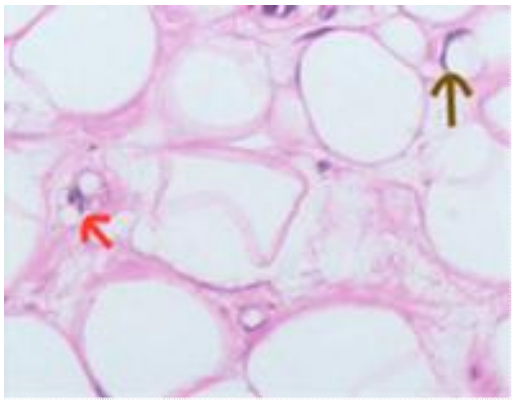


Figure 5: Atypical stromal cells in a myxoid background. H&E stain.
X40 magnification

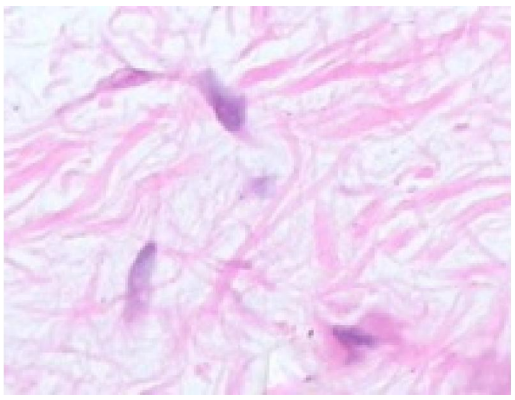
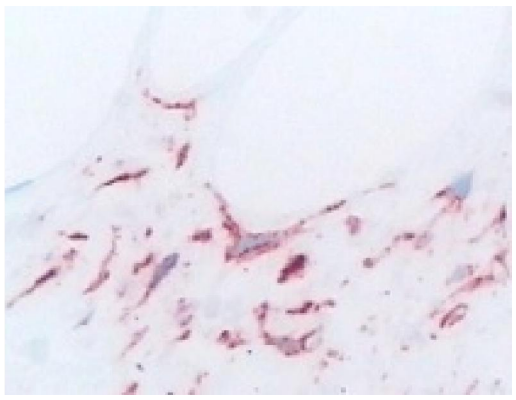


Figure 6: CD34 immunohistochemistry stain positive in the atypical stromal cells
X10 magnification



was 100% specific in the diagnosis of simple lipoma. The success rate of identifying well-differentiated liposarcomas was as follows: sensitivity, 100%; specificity, 83%; accuracy, 84%; positive predictive value, 38%; and negative predictive value, 100% [10].

Simple excision of a liposarcoma results in a considerably high local recurrence rate and may complicate further patient management. It is well known that malignant cells can penetrate a tumors' surrounding pseudocapsule, so simple removal of visible tumour in its capsular plane leaves microscopic disease behind. This likely contributes to the 90% recurrence rate after incomplete surgery without further treatment [4, 8]. In liposarcoma with positive histological margins, the disease will recur locally in more than 30% of cases even after additional excision of the tumour bed. Unfortunately, adjuvant radiotherapy does not compensate for positive histological margins after initially unplanned radical surgery [11]. Although there are no long-term outcome studies in this patient population, the prognosis depends on the treatment modality. Patients who receive only radiation therapy have significantly worse outcomes than patients who receive surgical resection or surgery plus adjuvant radiation therapy [1].

The importance of pre-operative planning, diagnostic core (tru-cut) needle, excisional or incisional biopsy has become essential in the algorithm for work up of high-risk soft-tissue tumours. This includes not only the previously described large and/or deep lesions, but also those that exhibit rapid increase in size or invasion of adjacent structures [12, 13].

Relative to this case, it would have yielded a better outcome if the fine needle aspiration cytology was performed earlier as per algorithm. Prior to surgery, we could have found as malignant cells in the FNAC, which could have prompted us to plan the further management more optimally.

CONCLUSION

Clinical suspicion and early detection, as well as treatment, should be advocated in all head and neck swellings. As discussed, accurate recognition and appropriate planning for the treatment will likely lead to a desirable outcome of better prognosis and disease-free resection margins. A missed step in the algorithm of head and neck swelling management, FNAC, resulted in a less than ideal management in this case.

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