Aggressive leiomyosarcoma of Pinna – A case report and literature review

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Abstract

In this article we present a rare aggressive case of a malignant mesenchymal tumour (leiomyosarcoma) presenting as a well circumscribed mass on the auricle. The tumour was treated with wide local excision and radiotherapy but unfortunately patient succumbed to the disease 24 months following initial presentation. We emphasize the importance of aggressive local control of this tumour at primary operation. Literature review of reported cases of leiomyosarcoma of the ear and its recommended clinical management are discussed.

INTRODUCTION

Leiomyosarcomas are malignant tumours arising from smooth muscle. These are rarely encountered in the head and neck region.[1,2] Though odd cases have been reported in nasopharynx, sino nasal tract and upper aero digestive tract, it's extremely rare in ears.[3,4,2,5] Only 9 cases of leiomyosarcoma involving ear have been reported in the literature (3 in auricle, 3 in external auditory canal, 2 in temporal bone and 1 in post auricular region).

CASE REPORT

A 78 year old man presented to us with 3 month history of fleshy nodular lesion on the right auricle. His past medical history include COPD, asthma and GERD. He smoked around 40 cigarettes a day for more than 50 years. Clinical examination revealed a 3x3 cms raised nodular, non tender lesion on the helical rim of right pinna, not attached to cartilage and without surrounding induration or regional lymphadenopathy. The patient underwent wide local excision aided by CO2 laser (4-6 watts) under local anaesthesia. Wound was closed primarily. At 2 weeks follow-up wound was healthy. The biopsy was reported as malignant mesenchymal tumour with positive immunochemistry staining for Vimentin and MSA, which is diagnostic of leiomyosarcoma. Margins were negative. The case was discussed at the regional cancer multi disciplinary meeting, where it was decided to regularly follow up the patient at short intervals. At 8 weeks follow-up the primary wound was healthy, but he had multiple small nodular swellings in the post auricular region. FNA of these confirmed metastasis. Within days he also developed a recurrent lesion of 1.5x1.5 cms anterior to the operated site at crus of helix and a node 2.5x 2.5cm in the post auricular region. CT scan of neck, chest and brain, USG abdomen were negative for any metastasis. His case was again discussed at the multi disciplinary meeting and as his general condition was fragile (COPD, oxygen dependence at home), decision for palliative radiotherapy was made. He received 60 grey in 25 fractions of external beam radiotherapy. Three months following radiotherapy post auricular nodule completely disappeared and the anterior one at root of helix was reduced in size. A regular monthly follow-up was organised. Six months latter he was admitted to the medical ward with shortness of breath, and subsequently was diagnosed with wedge fracture of lower thoracic vertebrae. Myeloma screen was negative.

At 18 months follow-up since presentation, his lesion on right ear started growing again and a 2nd course of palliative radiotherapy was given. He also developed a separate tiny crusty lesion on the left ear helical margin which was excised and biopsy was reported as squamous cell carcinoma with adequate margins. He ultimately succumbed to the metastatic leiomyosarcoma in his liver and bones 2 years after his initial diagnosis.

Figure 1

Fig 1: Leiomyosarcoma of Right pinna

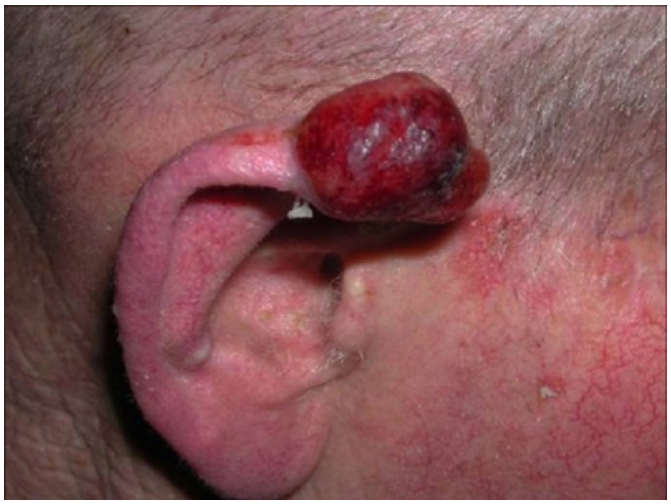


Figure 2

Fig 2: Excised specimen of pinna leiomyosarcoma

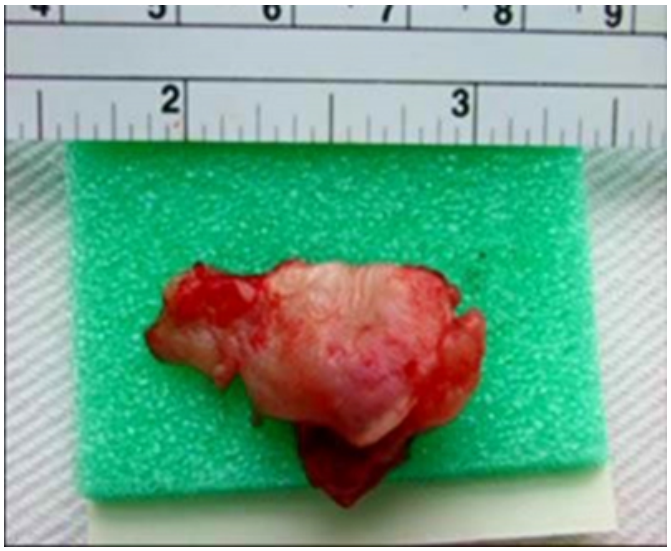
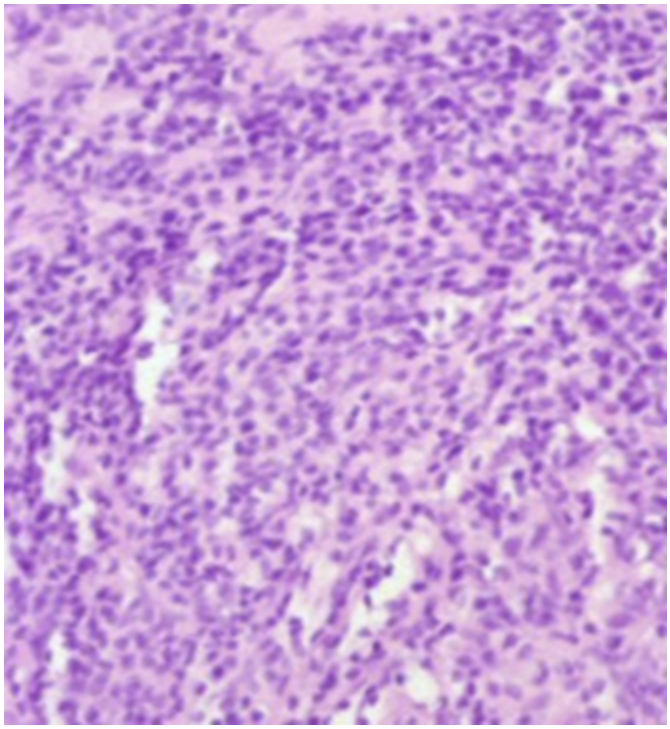


Figure 3

Fig 3: Histology photograph: malignant spindle cells arranged in fascicles



DISCUSSION

Leiomyoarcomas account for around 6% of all soft tissue sarcomas.[6] Head and neck region accounts for 3 to 10% of these tumours, found mainly in sinonasal tract, oral cavity and upper aero digestive tract.[3,4,2] Leiomyosarcoma of ear is extremely rare and only less than 10 cases have been reported in the literature.

As leiomyosarcoma is a tumour of smooth muscle cell origin, in ears the only possible origin is from the smooth muscle cells of local blood vessels or erector pili musculature of local hair follicles. As hair follicles are very sparse in auricle, in present case the most probable origin would be the smooth muscle cells of auricular blood vessels.

Mindell et al reviewed 31 cases of head & neck leiomyosarcomas and found most common site was scalp.[2] However Kuruvilla et al (9 cases) and Josephson et al (6 cases) in their series reported more commonly in sinonasal tract.[3,4] Detailed Pub med and Medline literature search revealed 9 cases reported in ears out of which only 3 involved the auricle. (Table 1)

Figure 4

Table: 1 - Reported cases of leiomyosarcoma of pinna and external auditory canal in the literature

S.No	Author	Journal	Year	Location
1	Oztürk K et al[7]	Auris Nasus Larynx.	2004	Post auricular region
2	Pai S et al[6]	Otolaryngol Head Neck Surg	2003	Auricle.
3	Karasen RM et al[1]	J Laryngol Otol	1998	Auricle
4	Nilles R et al[8]	Laryngorhinootologie	1995	Temporal bone
5	Zbären P et al[9]	Ann Otol Rhinol Laryngol	1994	Middle ear and temporal bone
6	Rasp et al[10]	Laryngorhinootologie	1991	External ear canal
7	Mindell RS[2]	Laryngoscope.	1975	Series of 31 cases including 1 in ear canal
8	Le Mouel C et al[11]	Rev Laryngol Otol Rhinol	1972	Auricle
9	Charlton C et al[12]	British J Surg	1964	External auditory canal

Leiomyosarcoma presents in the 5th to 6th decade of life and are twice more common in men than women.[7,6] They usually present as solitary nodules with intact overlying epidermis in the initial stages. They often metastasize by vascular routes to distant sites and lymphatic spread to regional lymph nodes is less common. Recurrence rate is between 40 and 60% and occurs generally during 1-5 years of primary treatment.[7,2] These most frequently metastasize to liver, lungs and bone by vascular spread. Mortality from subcutaneous leiomyosarcoma has been reported from 30 to 40%.[7,13] In one large series of leiomyosarcoma of head and neck, 20% were reported to have distant metastasis by haematogenous route and 10% had regional lymph node metastasis.[2] Overall survival rate ranged from 4 months to 12 years with mean survival of 2 years.[2] Our case had distant metastasis and succumbed to the disease after 2 years of initial diagnosis.

Light microscopic features of leiomyosarcoma are atypical spindle cells arranged in fascicles intersecting at right angles, intensely eosinophilic cytoplasm and centrally located cigar shaped nuclei. These poorly differentiated atypical spindle cells with blunted ends often blend into collagen. Immuno cytochemistry often establishes the diagnosis as these tumours stain positively for smooth muscle specific actin and vimentin. Differentiation of leiomyosarcoma from other spindle cell tumours like malignant schwannoma, neurilemmoma, spindle cell melanoma & spindle cell carcinoma etc is extremely difficult with light microscopy so immuno histochemical staining is essential for diagnosis.[7,1]

The mainstay of treatment for leiomyosarcoma is surgery i.e. wide local excision with adequate margins (1 to 2 cm), with wider margins for aggressive subcutaneous lesions. In head and neck, as its inherently difficult to establish wide resection margin, recurrence rates are often higher (30 to 50%). Lesions in the auricle can spread along the neurovascular bundle and embryonic fusion plates of the pinna, thereby invading surrounding structures (external auditory canal, temporal bone, parotid gland) early and making initial adequate resection difficult. Furthermore, relatively thin skin overlying the cartilage in pinna contribute to wider sub clinical horizontal spread along the dermis and pericondrium making initial judgement difficult.[6] Complete excision with safe oncological resection margin in this site includes total auriculectomy and total parotidectomy with radical neck dissection in the setting of regional metastasis. Elective neck dissection in clinically node negative disease (N0 Neck) is not recommended because of low incidence of lymphatic spread. As generally sarcomas are radio and chemo resistant tumours, the role of post operative radiotherapy and chemotherapy is controversial. In our case, radical surgical option was not available because of poor general condition of the patient and radiotherapy couldn't stop the aggressive tumour..

In conclusion, leiomyosarcomas are rare tumours in head and neck, need a high index of suspicion for diagnosis. Immunohistochemistry is essential for accurate diagnosis and surgery is the main stay of treatment. Even in pinna leiomyosarcoma can be aggressive enough to cause death if not treated with radical surgical excision.

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