Leiomyosarcoma Presenting as an Infected Pre-auricular Sinus

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Citation

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Abstract

Leiomyosarcoma in the head and neck region is rare. We describe a case in which a 44 year old man presents with an infected pre-auricular sinus. Histology of the lesion, however, revealed an unexpected diagnosis of leiomyosarcoma. A review of literature is presented.

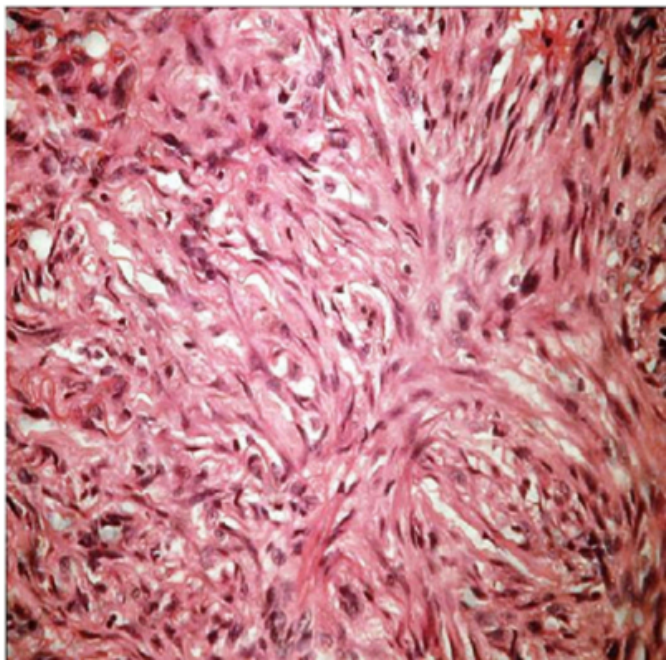
CASE REPORT

A 44 year old man is referred to the ENT clinic with a history of swelling in his left pre-auricular region for about 10-12 months. The swelling became inflamed intermittently. However the pre-auricular area was persistently tender. He received antibiotic treatment from his doctor during these episodes of inflammation. Examination revealed significant soft tissue swelling in the tragus, associated with tenderness. Anterior to the junction of the helix and tragus, an opening of a probable sinus tract was noticed with a small nodule below it. A provisional diagnosis of an infected pre-auricular sinus with perichondritis of the adjacent cartilage was made. Patient was treated with oral ciprofloxacin for a week. He was reviewed next week when no improvement in the symptoms recorded. Persistence of pain and tenderness even after antibiotic treatment raised the possibilities of other diagnosis including malignancies.

Frozen section of the lesion suggested sarcomatous changes and a wide excision of the lesion was carried out. Histology revealed a spindle cell lesion in the dermis with ill-formed fascicles and bland spindle cells focally admixed with histiocytic cells. The lesion was not well circumscribed and was invading the sub-mucus fat. Immuno-histochemistry showed the lesion to be desmin and HHF35 positive suggesting a tumor of smooth muscle origin, and melanoma and epithelial markers were found to be negative. A diagnosis of leiomyosarcoma was made.

Figure 1

Fig: Spindle cells arranged in fascicles. Nuclear atypia seen.



DISCUSSION

Leiomyosarcoma is a malignant neoplasm arising from smooth muscle. They constitute 4 - 6.5% of all soft tissue sarcomas [1, 2] with an overall incidence of approximately 0.04% among all cancers. [3] Leiomyosarcoma may occur at any anatomical site. In the head and neck area, smooth muscle is sparse and is found mainly in the walls of blood vessels and erector pili musculature of the skin. [4]

Cutaneous and subcutaneous leiomyosarcomas were formerly viewed as a single entity (superficial

leiomyosarcoma) but now are believed to be distinct diseases with dramatically different potentials for local recurrence and metastasis. Local recurrence rate for cutaneous leiomyosarcoma is only 30% compared to 70% for subcutaneous lesions. While subcutaneous lesions metastasize in 30-40% of the cases, distant metastasis is extremely rare in cutaneous leimyosarcomas. [4]

Superficial leiomyosarcoma is primarily a disease of middle age with a tendency to affect predominantly males. [5] Like other sarcomas, there is a predilection for extremities especially the lower extremities. Approximately 50 – 70% of the tumors involve lower limbs, 20 –30% upper limbs and 10% of these lesions are found in the trunk. [4] The incidence of facial tumors is around 1%. Pain is the most common symptom experienced in 80 – 95% of the patients. [5] Clinically, the lesions appear as single or multiple well circumscribed nodules.

Cutaneous leiomyosarcomas exhibit a fascicular, infiltrative peripheral growth pattern. Histopathologicaly, this neoplasm typically shows a proliferation of elongated spindle shaped cells in the dermis arranged partly in interweaving fascicles. The spindle cells show blunt – ended, cigar shaped nucleus with an eccentric vacuole, eosinophillic cytoplasm and conspicuous mitotic figures. [6, 7] Subcutaneous leimyosarcoma is usually surrounded by a compressed rim of connective tissue. Histologicaly, the lesion shows irregular aggregates of atypical myomatous spindle cells intertwined haphazardly. The distinct fascicular pattern that characterizes the cutaneous counterparts is not seen. Another conspicuous feature is the vascular pattern seen as endothelial-lined thin-walled vessels with lumens of differing dimensions and shapes. [5]

Pre-auricular sinus is a congenital lesion in which a small skin opening is located in front of the external ear communicating with a subcutaneous network of cysts. Majority of cases are benign in nature and require no intervention. However, recurrent infections in this seemingly trivial condition are not uncommon. This may lead to local scarring and secondary sinus formation. The treatment of choice in these cases is complete excision of the sinus tract and its associated cysts. Monie [8] noticed that in unilateral cases, left side is more frequently affected. It is interesting to note that in our patient, the lesion is on the left side as well.

Six cases of leiomyosarcoma of the ear have been reported. Of these, two were located in the external auditory canal [9,10] two in the auricle[8, 11] and two in the middle ear.[12,13] The treatment of choice of these lesions is wide local excision. Chemotherapy or Radiotherapy does not play any role in the treatment. [14]

The anatomical location of the sinus opening and the history of intermittent swelling are suggestive of a diagnosis of infected pre-auricular sinus. However, the presence of persistent pain, despite the antibiotic treatment in our patient raises the suspicion of other conditions. This case is reported to highlight the possibility of a rare diagnosis of a cutaneous leiomyosarcoma in patients with a clinical presentation mimicking an infected pre auricular sinus.

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