Med Discoveries
An open access journal of science and medicine

Article Type: Case Report
Volume 3, Issue 8
Received: Jul 10, 2024
Accepted: Aug 08, 2024
Published Online: Aug 15, 2024

Angiosarcoma of External Auditory Canal Masquerading as Pyogenic Granuloma

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Introduction

Sarcomas are rare and heterogeneous mesenchymal tumours that occur in deep soft tissue and bone. Most sarcomas are locally invasive with early metastatic potential since their progenitors are not delimited by basement membranes as are epithelial cells. Sarcomas are derived from mesoderm or neuroectoderm. They are classified according to type of tissue and resemble fat, fibrous tissue, smooth and skeletal muscle, endothelium, bone, cartilage, and nerves. Angiosarcomas are endothelial derived malignant neoplasms.

The diagnosis is often difficult, based on specific immunohistological features as they mimic benign lesions such as bruises or superficial infections. Hence, an early biopsy is mandatory. The mean survival time is 7 months if remains undiagnosed and untreated. This increases to 15 to 20 months in treated patients, 12 percent being alive at 5 years [1]. A dual therapy of surgery and radiotherapy offers the best chance of cure.

Here, we present a case of angiosarcoma masquerading as a pyogenic granuloma of the external auditory canal.

Case presentation

A 35-year-old male patient, driver by occupation presented to casualty with bleeding from right ear following trauma by goat horn since two weeks. The bleeding was intermittent, 1 teaspoon in quantity and stopped on applying pressure. It was associated with dull aching pain and decreased hearing since one week. There was no history of deviation of angle of mouth, vertigo, tinnitus or similar illness in past. On examination a shiny pink polypoidal mass 2 cm x 2 cm protruding from the external auditory canal of the right ear was seen, which was friable and bled on touch (Figure 1A). Tympanic membrane could not be visualised. Pure tone audiogram showed mild conductive hearing loss. Computed tomography showed well defined 0.85 cm x 0.72 cm x 1.62 cm soft tissue lesion filling the right external auditory canal extending upto tympanic membrane medially and mildly pushing it medially. No evidence of bony erosion was seen. No traumatic fracture line was noted in the temporal bone. Rest of the structures and ossicles appeared normal (Figure 1B). Excisional biopsy was planned through postauricular approach. Postaural Wilde's incision is taken and posterior meatotomy done. The canal is exposed. A reddish friable mass is seen arising from all around external auditory canal which is bleeds on touch, attached all around the external auditory canal skin. Removed in to and sent for histopathology. The external bony canal and tympanic membrane is found to be intact.

The histopathology report was suggestive of infected pyogenic granuloma and patient was discharged. However, the patient came back with multiple painless, erythematous, violaceous nodules (Figure 2) over the post aural scar and external auditory canal with perichondritis and preauricular inflammatory edema. Patient was admitted and treated for perichondritis. Resampling from postauricular lesion was done along. Resampling showed (Figure 3) vascular channels of varying size and shape with plump endothelium. Few of the vessels showed ectatic dilatation in cellular areas and solid areas. The endothelial cells revealed spindle like morphology with prominent mitotic figures and areas of haemorrhages and focal necrosis which were suggestive of angiosarcoma. Immunohistochemistry was done to confirm the diagnosis. IHC test showed CD 34 positive marker with 18-20% Ki-67 in highest proliferating area favouring diagnosis of low grade angiosarcoma.

Patient was referred for radiotherapy and chemotherapy. No further surgical intervention was done as patient refused surgery.

Citation: Gupta S, Kahane K, Jalvi M, Bardale M. Angiosarcoma of External Auditory Canal Masquerading as Pyogenic Granuloma. Med Discoveries. 2024; 3(8): 1199.

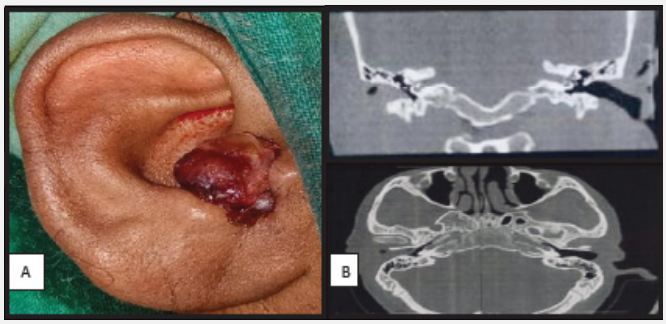

Figure 1: Clinical image.

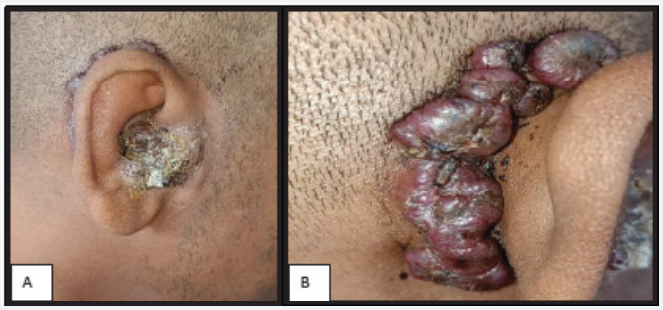

Figure 2: Painless, violaceous, nodules over the post aural scar and external auditory canal with perichondritis.

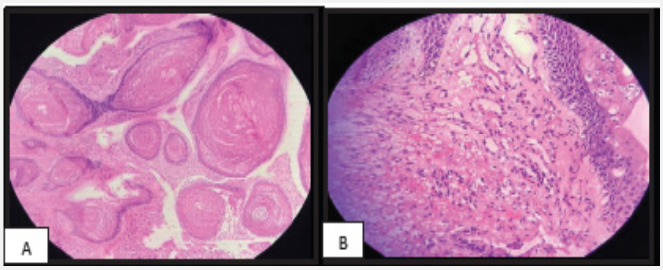

Figure 3: **A**: Histopathological slide shows collection of atypical endothelial cells forming keratin pearl like structures.
B: Histopathological slide showing anastomosing vascular channels.

Discussion

Angiosarcoma is a neoplasm of endothelial origin that arise most often in the skin, soft tissue, and viscera. It accounts for <2% of the sarcomas. Angiosarcoma of the ear is a very rare and locally aggressive malignant tumor. It accounts for 4-5% of cutaneous sarcomas and less than 1% of all sarcomas [2]. Men at an average age ranging from 60 to 80 years (sex ratio of 3:1) are most commonly affected with cutaneous angiosarcoma in head and neck region. The endothelial-derived neoplasms range from benign hemangiomas to tumours of intermediate malignancy such as hemangioendotheliomas to highly malignant and aggressive angiosarcoma. They arise from anywhere on the body involving the head and neck, particularly the scalp, followed by the breast, extremity, trunk, liver and other sites. Cutaneous angiosarcomas tend to arise spontaneously. Chronic lymphedema, a history of irradiation (Stewart-Treves syndrome) or environmental exposure to vinyl chloride and thorotrast contrast dye are well known causes of Angiosarcoma [3]. Genetic cancer mutations BRCA1, BRCA2, and NF-1 have also been implicated. Trauma has been identified as a risk factor for non- cranial angiosarcoma however the interval between the injury and diagnosis should be 1 month. Differential diagnosis of cutaneous angiosarcoma includes intravascular papillary endothelial hyperplasia, Kaposi sarcoma (multiple haemorrhagic sarcoma), Kaposi-like haemangioendothelioma, Angiolymphoid hyperplasia, Kimura's disease, Amelanotic melanoma, and Lymphangioma circumscriptum [4].

The prognosis for patients with angiosarcoma has generally been poor, as evidenced by a 5-year survival rate of 31%. The high incidence of advanced metastatic disease at the diagnosis (45% of cases) has likely contributed to the poor survival rate. However, those with localised disease at the initial diagnosis have had a 5-year survival rate of only 60%.

The diagnosis of angiosarcoma is dependent on the biopsy findings, with histologic features ranging from abnormal endothelial cells retaining some degree of well-differentiated vascular architecture to poorly differentiated sheets of abnormal cells with significant haemorrhage and necrosis. Immunohistochemistry is the gold standard test for Angiosarcoma which gives positive staining for CD31, CD34, factor VIII-related antigen, and ERG [5]. Computed tomography is useful to evaluate the tumour characteristics and architecture and for accurate staging of the tumour with invasion of the soft tissues.

There are no definitive surgical guidelines for the management of cutaneous disease. The French Society of Otorhinolaryngology guidelines regarding the management of cutaneous angiosarcomas involves performing the widest possible resection (2 to 5 cm margins). However, complete resection is not possible in most circumstances as a result of diffuse, noncontiguous tumour margins and significant structural and functional impairment. 40% of angiosarcomas are metastatic on presentation, further restricting the therapeutic efficacy of surgical resection. There are multiple adjuvant therapeutic modalities. Adjuvant radiotherapy has been a mainstay of therapy. Adjuvant chemotherapy can be done with traditional agents like taxanes and doxorubicin, bevacizumab, TRC105, trebananib and the vascular endothelial growth factor receptor inhibitors pazopanib, sorafenib, axitinib and phosphoinositide dependent protein kinase 1. β-blockers also have a potential role in treating angiosarcoma.

At present, the treatment of angiosarcoma is often individualised, and even the optimal use of all available modalities has been associated with transient responses and a poor prognosis. Because of the strong metastatic potential, an extensive staging work up should be performed in all cases at the time of diagnosis.

Conclusion

Angiosarcoma is a rare tumour. Due to its nonspecific early signs, diagnosis can be delayed. Histopathological examination and Immunohistochemistry are the keys to the diagnosis and effective treatment. Age, tumour size, and disease extent are determinants of head and neck cutaneous Angiosarcoma survival. There is possibility of late local recurrence or distant metastasis, hence lifelong follow-up is necessary.

Keywords: Brucellosis; Human; Sero-prevalence; Small ruminant Ethiopia.

Declarations

Source of financial support or funding: Nil.

Conflict of interest: Nil.

Ethical consideration: Informed written consent were taken from the patient for taking photographs and publication purposes. Permission was taken from the ethical committee for publication purpose. No harm was done to animals for this study.

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