Paediatric ALK-Positive Anaplastic Large Cell Lymphoma of External Auditory Canal: A Rare Differential Diagnosis

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ABSTRACT

Anaplastic large cell lymphoma (ALCL), ALK-positive, is an uncommon disease. It is an aggressive CD30-positive T-cell lymphoma with a chromosomal rearrangement and expresses the ALK gene and ALK protein. Malignancies in the head and neck region rarely occur in the ear. Hence, the initial presentation of ALCL in the ear frequently leads to misinterpretation and difficult diagnosis. We present a case of a seven-year boy who presented with a polypoidal mass in his left ear and was treated as a case of otitis media for some time but was eventually diagnosed as a case of ALK-positive ALCL, small cell variant.

Key Words: *ALK-positive lymphoma, Facial palsy, Otitis media, Non-Hodgkin lymphoma.*

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INTRODUCTION

Lymphomas are the most prevalent non-epithelial tumours in the head and neck region and the third most frequent malignancy overall. About 5 to 15% of head and neck tumours are lymphomas; of which, 60% are non-Hodgkin lymphomas (NHLs), while 40% are Hodgkin lymphomas (HLs).[1] Among the types of NHL, anaplastic lymphoma kinase (ALK)-positive, anaplastic large cell lymphoma (ALCL) is a relatively rare kind that is an aggressive CD30-positive T-cell lymphoma with a chromosomal rearrangement expressing the ALK gene and ALK protein.[2] ALK-positive ALCL is further divided into five morphological patterns: The common pattern, the lymphohistiocytic pattern, the small cell pattern, the Hodgkin-like pattern, and the composite pattern.[3] We, herein, discuss an instance of an external auditory canal mass in a child, which was later on diagnosed to be ALK-positive ALCL, a small cell variant.

CASE REPORT

A seven-year boy with complaints of a reddish mass coming out of the left ear and swelling behind the left ear visited the otorhinolaryngology department. The mass was rapidly enlarging and was not associated with discharge and pain. The swelling was sudden in onset and painful and associated with fever and headache. It is worth noting that there was no complaint of facial weakness, tinnitus or vertigo, double or decreased vision, dizziness, or neck stiffness at the first presentation.

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Examination revealed a red polypoidal mass coming out of the left ear which bled on probing. The swelling behind the left ear was 3×3 cm in size, tender, smooth, and had well-defined margins. It was soft and fluctuant in consistency. The overlying skin was inflamed and there was a punctum. It was not fixed, non-pulsatile, non-reducible, with no transillumination (Figure 1). The tuning fork test showed negative Rinne's test in the left ear and positive in the right ear while the Weber lateralised towards the left ear. Initially, injectable antibiotics were started, and the patient's signs and symptoms started to improve. The patient was discharged on oral broad-spectrum antibiotics.

One month later, the child presented with worsening symptoms and facial nerve paralysis (Figure 2) and an ulcerated lesion on the hard palate.

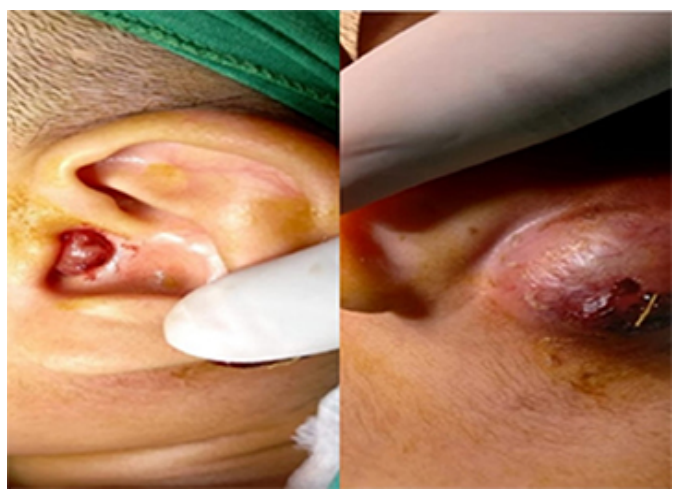

Figure 1: Aural polyp in the left external auditory canal and the post-auricular swelling on the same side.

Table I: All cases of ALCL of the external auditory canal reported from 2000 – 2023.

Authors	Year	Age	Gender	Location	Presenting complaint	Treatment	Facial nerve palsy	Other locations on PET CT	ALK status
Merkus *et al.*[6]	2000	83 year	Female	Lt. External auditory canal	Otalgia and otorrhea	RT	Yes	No	NA
Gomaa *et al.*[7]	2010	47 year	Female	Lt. External auditory canal	External auditory canal swelling	RT	NA	Lt. Side level II cervical lymph node	NA
Marcal *et al.*[4]	2012	68 year	Female	Rt. External auditory canal	Otalgia and hypoacusis	RT	NA	No	-VE
Rodriguez *et al.*[8]	2012	6 year	Male	Rt. Ear lobule	Rt. Ear lobule swelling	Surgical excision	No	No	-VE
Chen *et al.*[9]	2021	34 year	Male	Lt. External auditory canal	Painless mass and otorrhea	CCRT	No	No	+VE
Our study	2023	7 year	Male	Lt. External auditory canal	External auditory canal mass and postauricular swelling	CCRT	Yes	Liver, para-aortic and aortocaval lymph node	+VE

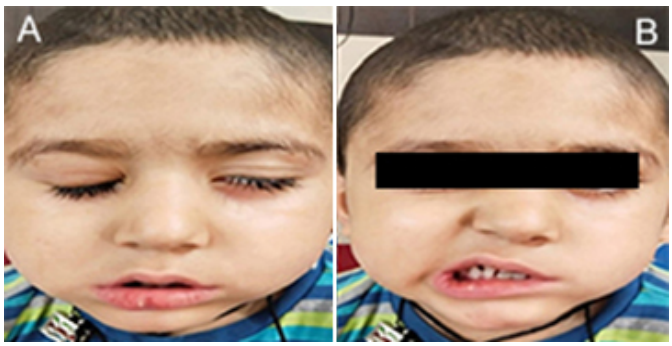

Figure 2: Facial paralysis. (A) Incomplete eye closure. (B) Deviation of the angle of the mouth.

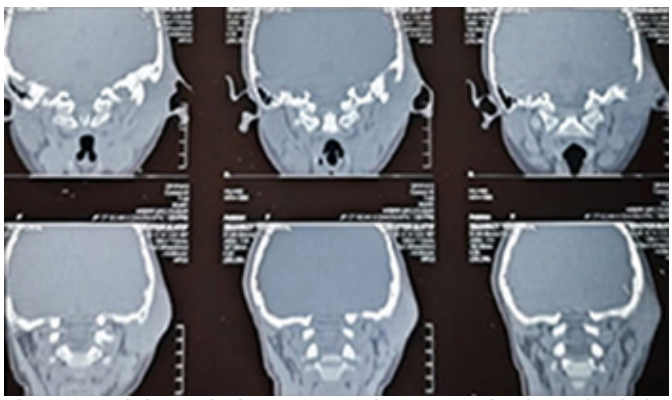

Figure 3: High-resolution computed tomography (HRCT) of the temporal bone.

High-resolution computed tomography (HRCT) of temporal bone identified poorly defined heterogeneous density mass along left mastoid air cells causing their complete opacification, with the erosion of lateral wall and extension of the soft tissue component in adjacent periauricular and retro-auricular space. It was also extended into the external auditory canal causing its complete opacification up to the level of the tympanic membrane. The sinodural plate on the left side also appeared partially eroded (Figure 3). MRI revealed an ill-defined area of abnormal signal intensity in the left middle ear extending into the left retro-auricular region and inferiorly along the posterior aspect of the superficial lobe of the left parotid and encasing the left facial nerve. Intracranial extension with dural enhancement was also noted as well as a small nodular intracranial enhancing lesion along the left cerebellar hemisphere's inferior surface (Figure 4).

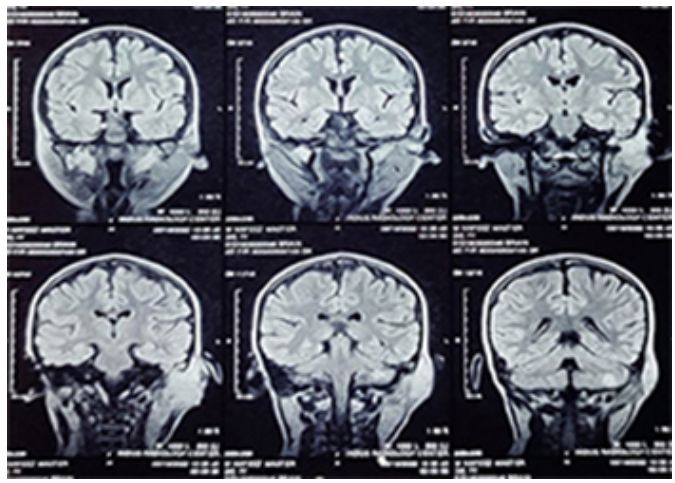

Figure 4: MRI brain with gadolinium enhancement (coronal view).

Multiple biopsies of the mass were taken and sent for histopathology. The histological assessment showed extensive lymphoid infiltrates comprising of small cells having plasmacytoid appearance with round to oval hyperchromatic nuclei and scant pale cytoplasm. Immunohistochemical (IHC) analyses were positive for leucocyte common antigen (LCA), CD3, CD30, ALK, EMA, and CD4, and the Ki-67 proliferative index was 80%. The findings were consistent with ALK-positive ALCL, a small cell variant. Further workup included a CT scan of the chest, abdomen, and pelvis, and a bone marrow biopsy which displayed large para-aortic, aorto-caval, and bilateral iliac lymph nodes, and normal trilineage haematopoiesis, respectively. Therefore, based on the Ann-Arbor staging system of primary systemic ALCL, the patient was staged as stage IV.

The patient was immediately referred to the oncology unit, where the patient was started on chemotherapy with Bleomycin, Methotrexate, and Venorelbine (BMV) regimen. At the latest follow-up, after four months of diagnosis and five cycles of chemotherapy, the patient had stable control of the disease and his facial nerve palsy had also reversed.

DISCUSSION

ALK-positive ALCL is a subtype of peripheral T-cell lymphoma (PTCL) characterised by large lymphoid cells with copious

amounts of cytoplasm and pleomorphic, frequently horseshoe-shaped nuclei, high CD30 immunostaining, and ALK chromosomal translocation.[4] This lymphoma usually affects extra nodal as well as nodal locations. Skin, bone, soft tissue, lung, and liver are the extranodal areas that are most frequently affected. At initial diagnosis, most patients have stage III or IV constitutional symptoms. When using standard haematoxylin and eosin (H and E) stains for analysis, the incidence of bone marrow involvement is roughly 10–11%, but when ancillary IHC tests are performed, the detection rate rises to 30%.[5]

On histologic examination, the dermal involvement in primary T-cell lymphoma lesions is either nodular and/or diffuse, and it frequently extends to the subcutis. Large anaplastic cells normally make up the cell population which are arranged in broad, confluent sheets that are cohesive. The histologic criteria for a primary cutaneous CD30-positive T-cell lymphoma diagnosis include the presence of CD30 positivity in no less than 75% of the lymphoma cells.[4]

In this case, the patient presented with an ear mass. The differentials included in this case were chronic suppurative otitis media with aural polyp, foreign-body granuloma, otitis externa, haemangioma, squamous cell carcinoma, and primary T- cell lymphomas. However, it is worth noting that only a biopsy can reveal the exact nature of these types of tumours, because they all initially appear as non-specific cutaneous disease.

To the best of our knowledge, only a handful of cases have been reported of ALCL of the external auditory canal, as shown in Table I.[6-9]

Therefore, based on the literature search, this case is the first instance of ALK-positive ALCL (small cell variant), and the second case of ALK-positive ALCL in the external auditory canal that has been reported in childhood age group.

The small cell variant of the ALK-positive systemic ALCL has been reported to involve the peripheral circulation more frequently. Compared to the other types, they change into and from the common variant of ALCL and have a worse prognosis overall. According to reports, patients with the small cell variant of ALCL had a 50% two-year survival rate, compared to 73% for patients with the common type. As mentioned above, certain cases of the small cell variant of ALCL are quite aggressive despite being ALK-positive. It is unclear whether this is due to the dispersed nature of the tumour in many patients or to tumour biology that is more aggressive.[10]

The small cell variant of ALCL is an uncommon disease that poses a diagnostic difficulty and is harder to identify. When a young patient presents with a soft tissue mass in the external auditory canal and postauricular swelling with an aggressive pattern of growth and is resistant to conventional medical treatment, otolaryngologists should consider this entity in the differential diagnosis, especially in a case of a chronic suppurative otitis media with aural polyp.

PATIENT'S CONSENT:
Consent was obtained from the patient's parents to publish this case report.

COMPETING INTEREST:
The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
SFA, AJ, SZ: Designing, drafting, and critical revision of the manuscript.
DUR: Literature review.
IAS: Proofreading.
All authors approved the final version of the manuscript to be published.

REFERENCES

1. Huyett P, Johnson JT. Extranodal lymphoma of the posterior auricle. *Am J Otolaryngol* 2015; **36(4)**:587-9. doi: 10.1016/j.amjoto.2015.03.005.
2. Ferreri AJ, Govi S, Pileri SA, Savage KJ. Anaplastic large cell lymphoma, ALK-positive. *Crit Rev Oncol Hematol* 2012; **83(2)**:293-302. doi: 10.1016/j.critrevonc.2012.02.005.
3. Montes-Mojarro IA, Steinhilber J, Bonzheim I, Quintanilla-Martinez L, Fend F. The pathological spectrum of systemic anaplastic large cell lymphoma (ALCL). *Cancers* 2018; **10(4)**:107. doi: 10.3390/cancers10040107.
4. Marcal N, Campelos S, Dias L, Goncalves M, Pereira G, Godinho T. Primary cutaneous CD30-positive anaplastic large-cell lymphoma of the external auditory canal. *Ear Nose Throat J* 2012; **91(12)**:E10-2. doi: 10.1177/014556131209101214.
5. Pasricha S, Gandhi JS, Gupta G, Mehta A. Small cell variant of anaplastic large cell lymphoma presenting as arm mass in a child: A rare entity with diagnostic challenge. *J Cancer Res Ther* 2013; **9(2)**:317-9. doi: 10.4103/0973-1482.113410.
6. Merkus P, Copper MP, Van Oers MH, Schouwenburg PF. Lymphoma in the ear. *ORL J Otorhinolaryngol Relat Spec* 2000; **62(5)**:274-7. doi: 10.1159/000027759.
7. Gomaa O, Serafini AN, Franzmann E, Kittaneh M, Montague N, Kalkanis D. F-18 FDG PET/CT in the assessment of recurrent anaplastic T-cell lymphoma of the external auditory canal. *Clin Nucl Med* 2010; **35(8)**:592-4. doi: 10.1097/RLU.0b013e3181e4db98.
8. Rodriguez V, Perolada JM, Ibanez I, Fernandez A. Anaplastic large T-cell lymphoma of the external ear in childhood. *Acta Otorrinolaringol Esp* 2013; **64(3)**:230-2. doi: 10.1016/j.otorri.2012.01.004.

9. Chen SL, Chan KC. Anaplastic lymphoma kinase- and CD30-positive anaplastic large-cell lymphoma of the external auditory canal. *Diagnostics (Basel)* 2021; **11(7)**:1220. doi: 10.3390/diagnostics11071220.

10. Summers TA, Moncur JT. The small cell variant of anaplastic large cell lymphoma. *Arch Pathol Lab Med* 2010; **134(11)**:1706-10. doi: 10.5858/2008-0624-RSR.1.