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CASE REPORT

Infantile Otorrhea: Not As Benign As Perceived

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Key words: Otorrhea; Temporal Bone Tumour; Ewing Sarcoma

ABSTRACT

Otorrhea in preschool children is most often infective and benign in nature. Temporal bone tumours presenting with persistent otorrhea often have associated features such as raised intracranial pressure, temporal bone swelling and facial nerve palsy. We describe an infant with temporal bone Ewing sarcoma presenting with isolated chronic otorrhea and review the clinical features of the four commonest paediatric temporal bone tumours over the last 50 years.

Keywords: Otorrhea; Temporal bone tumour; Ewing sarcoma;

Case Report

A 9-month-old boy was referred for concerns of recurrent foul-smelling discharge of the right ear for 1 month. At first presentation, his family physician diagnosed a right otitis externa with microabscesses and commenced oral Ampicillin/Sulbactam with topical ofloxacin. The ear discharge resolved within a week but recurred again three days prior to presentation to us. It was yellow in colour, foul-smelling and with no associated bleeding. His appetite, behavior, and sleep were normal with no indication of pain or distress. No associated URTI, fever or constitutional symptoms.

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He was born term via spontaneous vaginal delivery with a birth weight of 3kg with an uneventful postnatal period. Vaccinations completed for age. On examination, he was afebrile, fretful when approached, with no facial asymmetry or focal neurological deficits. A subtle non-tender swelling of the right mastoid region was noted, with minimal displacement of the right pinna anteriorly. No temporal region or peri-auricular swelling seen and no palpable lymphadenopathy. Cranial nerve examination, including facial nerve examination, was normal. Pupillary reflexes were equal on both sides. Otoscopic examination of the right ear revealed bulging of the posterior external auditory canal (EAC) (Figure 1a) with purulent pus discharge and inability to pass the speculum beyond the swelling to visualise the tympanic membrane. Left ear examination was normal. An initial diagnosis of cholesteatoma was suspected. Brainstem evoked response (BSER) confirmed right profound sensorineural hearing loss with normal hearing in the left ear.

Examination under anesthesia of the right ear revealed extensive granulation tissue filling the entire EAC from lateral to medial with posterior canal wall erosion and the tympanic membrane could not be visualised (Figure 1b).

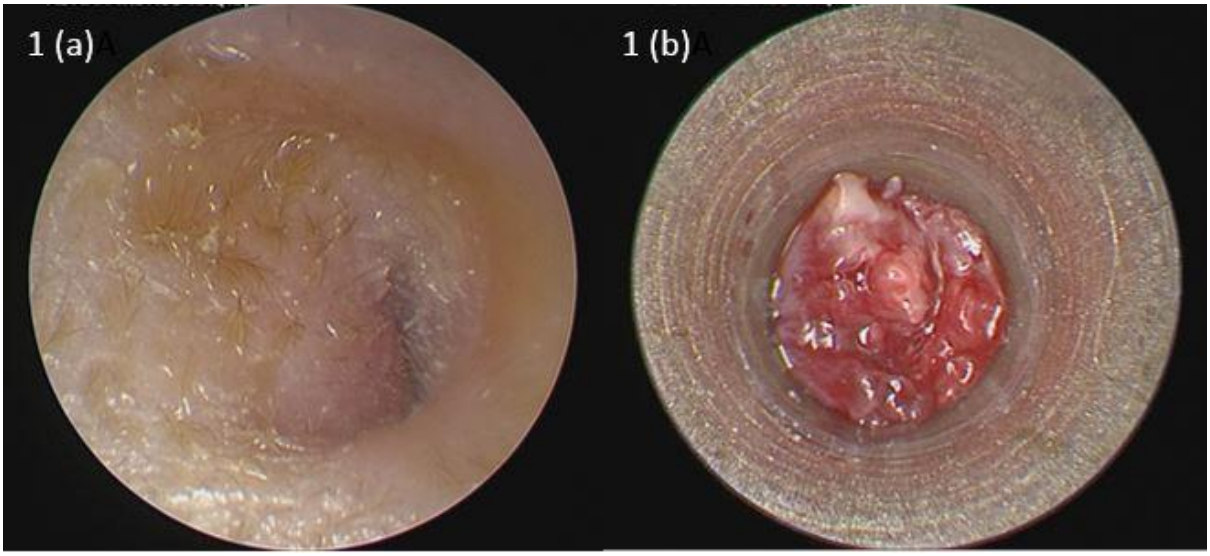

Figure 1: Intraoperative view of the EAC showing bulging of the posterior EAC occluding the ear canal (1a). Tumour filling the entire EAC (1b).

CT scan of temporal bone showed patchy expansile destruction of right mastoid bone with soft tissue components measuring 3.0 x 4.0 x 3.2cm with intracranial extension into the adjacent temporal and cerebellum (Figure 2).

MRI brain revealed a huge expansive destructive mass measuring 4.3 x 4 x 3.5cm within the right mastoid and petrous with vividly enhancing leptomeningeal component and partial involvement of the right temporomandibular joint (Figure 3).

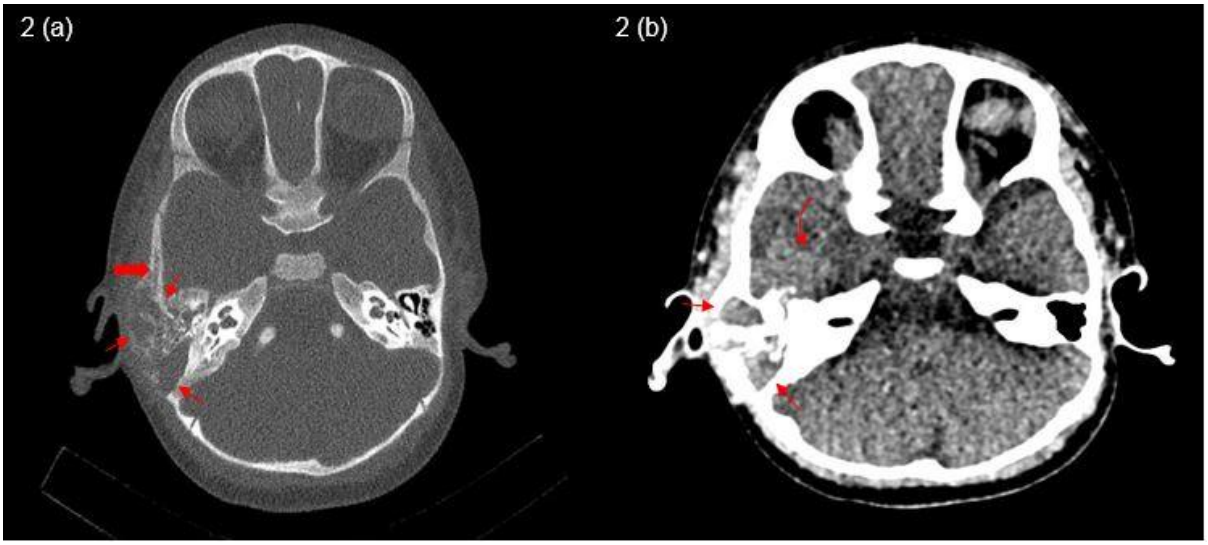

Figure 2: Non-contrasted bone (2a) & soft tissue window (2b) images of temporal bone CT scan show expansile bony lytic destructive lesion with soft tissue component within (thin red arrow) involving the mastoid portion of right temporal bone with periosteal reaction (thick red arrow) and intracranial extension (curved red arrow) into regional temporal lobe.

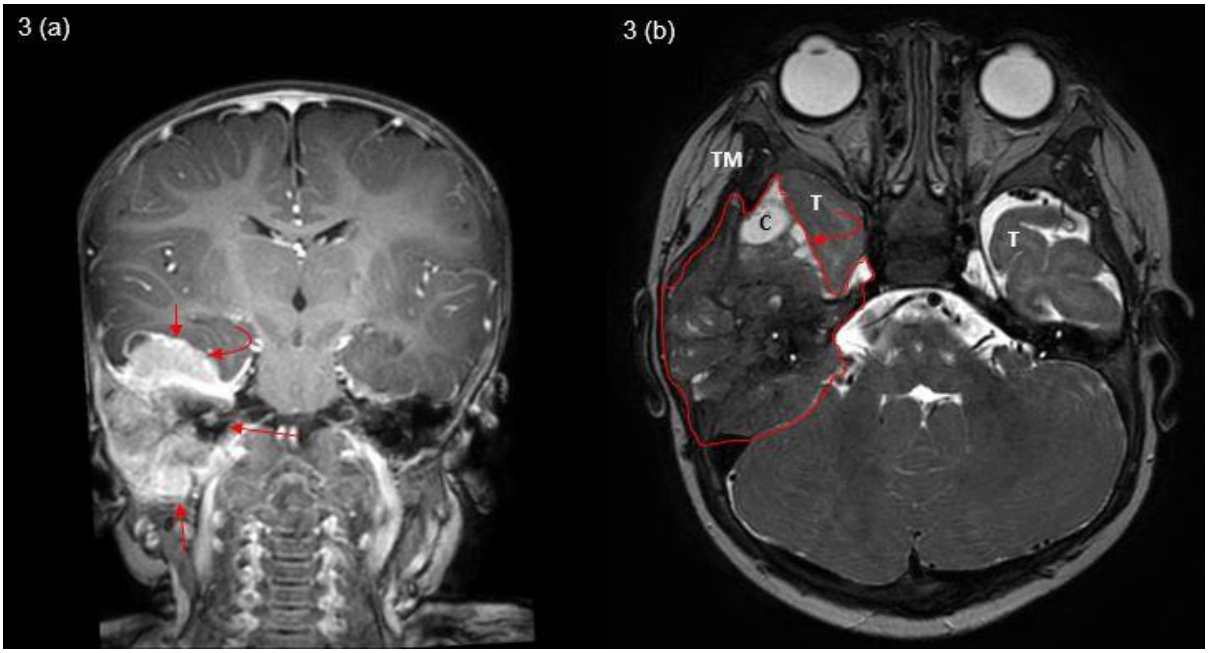

Figure 3: T1-weighted MRI with gadolinium (3a) and T2-weighted (3b) images of the brain showing an expansile (outlined) destructive heterogeneous intensely enhancing mass (arrow) adjacent to the right temporalis muscle (TM) and involving the mastoid as well as petrous bones with a small cystic component (C) at anterior aspect and intracranial extension into regional temporal lobe (T) (curved arrow).

Intraoperatively, the granulation tissue was sent for microbiological studies which showed no growth. Histopathological analysis revealed fragments of loose fibromyxoid tissue with infiltrating sheets of small round cells with indistinct cytoplasm and occasional mitosis, which was suggestive of a small round cell tumour. The cytogenetic report was consistent with Ewing sarcoma. Fluorescence in situ hybridization (FISH) analysis showed an abnormal result with evidence of EWSR1 (22q12) rearrangement in 118 out of 200 (59%) interphase nuclei scored. EWSR1 rearrangements are recurrent, non-random abnormalities associated with Ewing Sarcomas. Approximately 90% of Ewing tumours show a t(11;22) (q24;q12). This translocation results in the fusion of the EWSR1 gene at 22q12 with the transcription factor gene FLI at 11q24, leading to an oncogenic chimeric protein (Figure 4).

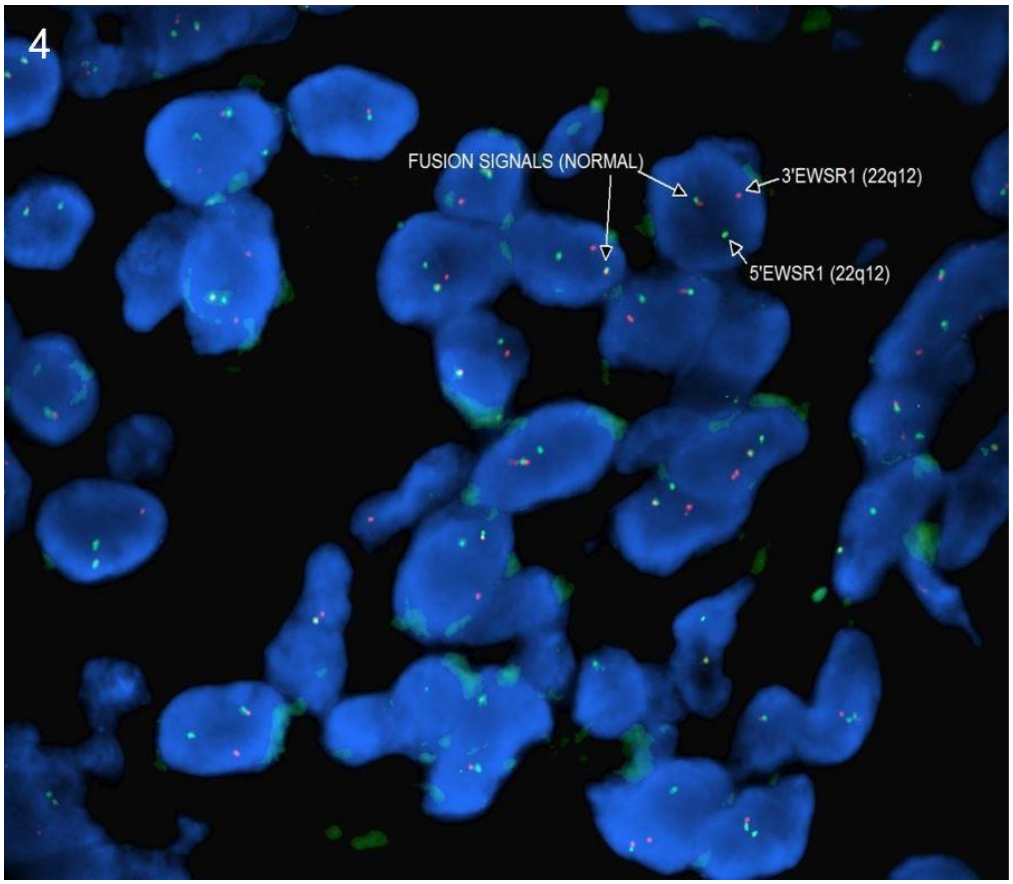

Figure 4: FISH Positive for Ewing Sarcoma with split signals in most nuclei. EWSR1 (22q12) gene rearrangement pattern observed by a split-signal probe showing a pair of fused (yellow) and split (separated red and green) signals.

The child received 6 cycles of chemotherapy which consisted of the VIDE regime: Vincristine, Ifosfamide, Doxorubicin, and Etoposide. Post-treatment MRI scans showed a reduction in tumour size, especially the intracranial component, with tumour size of 2.7 x 3.7 x 2.1cm, with persistent dural enhancement. Following a multi-disciplinary team discussion, the child was listed for right radical mastoidectomy and excision of dura. Due to Salmonella infection, the surgery was postponed

DISCUSSION

Otorrhea arises as a result of disease from either the external auditory canal or middle ear, following drainage from a perforated tympanic membrane. Though the commonest cause of otorrhea in preschool children is acute and recurrent otitis media[1], other recognised causes of importance include neoplasms, cholesteatoma, dermatitis, otomycosis and foreign body obstruction within the auditory canal. Most often, an underlying infective cause is assumed and antibiotics prescribed. If treatment failure ensues, further evaluation by an otolaryngologist is advised.

Malignancy of the temporal bone is a rare entity, with an incidence rate of 0.2% of all head and neck cancer in the general population[4]. The incidence of temporal bone tumours in children remains obscure due to its rarity. Various types of paediatric temporal bone tumours have been recognised including rhabdomyosarcoma (RMS), Langerhans cell histiocytosis (LCH), osteosarcoma (OS), Ewing sarcoma (ES), chondrosarcoma, lymphoma, and fibrosarcoma. Of these, the commonest are RMS and LCH[3].

Primary ES is a small round cell tumour of neuroectodermal origin. It is the second most common primary bone tumour in children and accounts for approximately 8% of all malignant bone tumours in children. Primary ES most commonly affects the truncal and long bones and rarely the skull base (1% of all Ewing's sarcoma)[5,6].

Most often, temporal bone tumours have non-specific signs and symptoms, leading to delayed diagnosis. Our patient presented only with recurrent painless otorrhea and a subtle mastoid swelling with no other focal neurology, despite significant intracranial and extracranial tumour extension. Important clues that can help differentiate benign otorrhea from a more sinister cause include the presence of associated hearing loss, otalgia, tinnitus, extra auricular mass, facial nerve palsy, blurring of vision, diplopia, proptosis, ophthalmoplegia, temporal, mastoid, or peri-auricular swelling and ataxia.

Table 1: Clinical features based on histological types of four commonest temporal bone tumours in children over the last 50 years (n= 105)

Clinical features \ Tumour types	ES, *n*=28	RMS, *n* = 25	LCH, *n* = 49	OS *n* = 3
	n (%)	n (%)	n (%)	n (%)
Otorrhea	1 (3.57%)	17 (68%)	22 (44.9%)	1 (33.33%)
Extra-auricular mass		10 (40%)	3 (6.12%)	
Hearing loss	4 (14.29%)	7 (28%)	3 (6.12%)	
Otalgia	1 (3.57%)	8 (32%)	11 (22.45%)	
Tinnitus	1 (3.57%)			
Raised ICP*	23 (82.14%)			
Facial nerve palsy	9 (32.14%)	15 (60%)	1 (2.04%)	
Temporal region swelling	14 (50%)	2 (8%)	1 (2.04%)	
Mastoid swelling	1 (3.57%)		2 (4.08%)	
Preauricular swelling		2 (8%)	18 (36.73%)	2 (66.67%)
Parotid swelling		1 (4%)		
Ocular#	4 (14.29%)	3 (12%)	3 (6.12%)	
Others+	2 (7.14%)	3 (12%)	4 (8.16%)	

* Raised ICP: vomiting, headache, drowsiness.
# Ocular: blurring of vision, diplopia, ophthalmoplegia, proptosis, eye edema.
+ Others: lymphadenopathy, seizure, ataxia, seborrhea

Table 1 illustrates the symptoms and signs of one hundred and five children (5 months to 19 years old) with temporal bone tumours reviewed over the last 50 years.

RMS and LCH present most commonly with otorrhea mimicking otitis media. Facial nerve palsy (60%) is the next commonest associated symptom in RMS, whereas peri-auricular swelling (36.7%) is the second most common symptom presentation in LCH.

Symptoms of raised intracranial pressure (82.14%) are the most frequent presentation in ES. Temporal bone swelling and facial nerve palsy are the next commonest presenting features of ES with otorrhea being the least frequent presenting complaint reported in only one case, with ours being only the second case published to date. Owing to the rarity and varied presentations of temporal bone tumours in children, differentiating histological types based on symptoms alone appears difficult.

RMS and LCH however, appear to be the commonest temporal bone tumours seen and should be an important differential diagnosis to exclude, in children with non-resolving otorrhea particularly if associated with facial nerve palsy and peri-auricular swelling. Raised ICP surprisingly was not a dominant symptom seen in either two of these temporal bone tumours but was the most common presentation in ES. The presence of an expansile mass with intracranial extension at presentation, in the majority of cases of skull ES, could explain this unique observation[7].

CT imaging of the temporal bone has shown to help delineate the integrity of the bony structures accurately. Further, the ossicles, skull base, bony labyrinth, vestibule, internal auditory canal, mastoid air cells, fallopian/facial nerve canal, eustachian tube and petrous apex can be evaluated. Conversely, magnetic resonance imaging (MRI) does not display bony destruction as well as CT scan but provides significantly higher resolution for delineating lesions infiltrating the brain, parotid space, masticator space, parapharyngeal space, infratemporal fossa and subcutaneous tissue[3]. As in our patient, the expansile destructive mass of the temporal bone visualised on CT imaging is in keeping with the radiographic description of ES in existing literature.

Though outcomes of ES remain complex, the prognosis of ES of the head and neck region appears more favourable than extracranial ES, owing to the reduced risk of distant metastasis. In a recent review, only one reported case of cranial ES displayed distant metastasis during follow-up period[6].

CONCLUSION

The presence of chronic otorrhea in preschool children may necessitate evaluation by an otolaryngologist to rule out associated serious pathologies. The diagnosis of temporal bone tumours is often delayed as the presenting signs and symptoms are usually non-specific. Awareness of the association between temporal bone tumours and recurrent otorrhea with associated pathological signs in children is vital. Early diagnosis and treatment before metastasis have shown to improve long-term survival in patients with Ewing sarcoma.

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