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Case Report

Pathology Section

Embryonal Rhabdomyosarcoma in Middle Ear: A Rare Case Report

ANITA OMHARE[1], SANJEEV KUMAR SINGH[2], GEETA MAURYA[3], ROOPAK AGGARWAL[4], SANJAY KANNAUJIA[5]

ABSTRACT

Rhabdomyosarcoma (RMS) in middle ear and mastoid are uncommon. Presentation of RMS in these sites include purulent or blood stained discharge, polypoidal mass and granulation tissue. Clinical diagnosis may be delayed due to similar presentation as Chronic Suppurative Otitis Media (CSOM). Authors herein report a case of six-year-old male patient, who presented with a blood stained discharge from right ear for 15 days. On otoscopic examination, a polypoidal mass in external auditory canal was identified. A history of ruptured right tympanic membrane with occasional discharge for two years had been given. Clinical diagnosis of CSOM with aural polyp had been made. Radiographically, it was a soft tissue lesion with poorly defined margins. There were no bony erosions with minimal collection in right middle ear cavity. On routine histopathological examination diagnosis of round cell tumour had been made. On Immunohistochemistry (IHC), tumour was immunoreactive for desmin and vimentin, while negative for Cytokeratin (CK), Leukocyte Common Antigen (LCA), chromogranin and synaptophysin. Tumour was diagnosed as RMS (Embryonal type). This case report shows importance of early diagnosis of RMS in middle ear, which is often delayed and misdiagnosed as CSOM. Delayed diagnosis may lead to, facial nerve involvement, local meningeal involvement and distant metastasis. Histopathological examination and IHC e.g., desmin positivity are the mainstay of diagnosis.

Keywords: Desmin, Immunohistochemistry, Otitis media, Round cell tumour

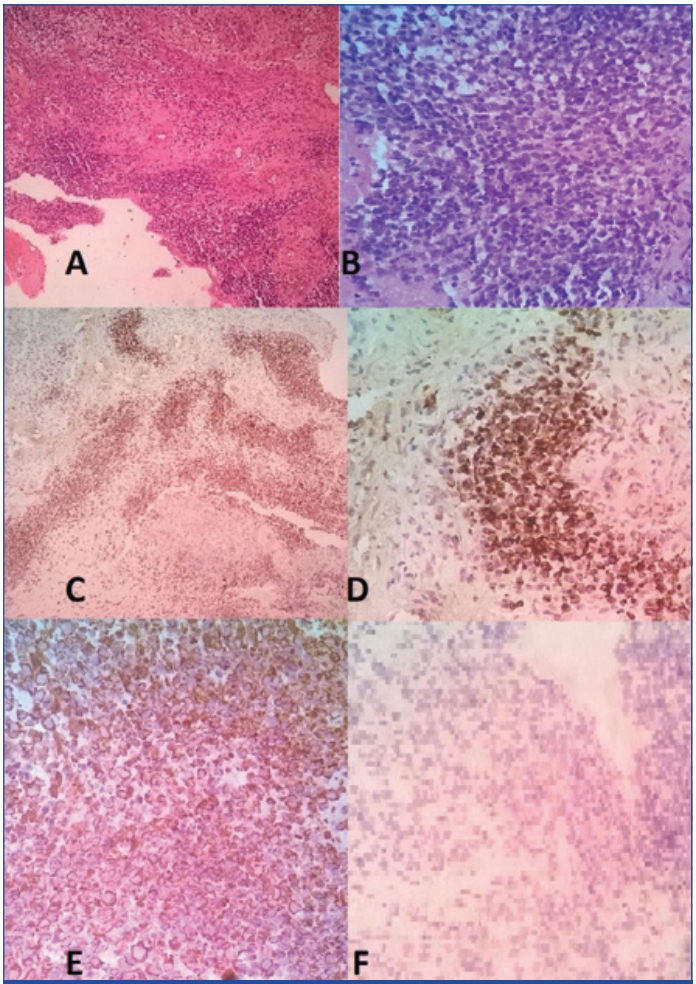
CASE REPORT

A six-year-old male patient reported to Outpatient Department of Otorhinolaryngology with a chief complaint of blood stained discharge from right ear since 15 days. Patient gave history of ruptured right ear tympanic membrane two years back and since then patient noticed occasional serosanguineous discharge from right ear. On otoscopic examination, a polypoidal mass in external auditory canal was identified. There was minimal collection in right middle ear cavity. Clinical diagnosis of CSOM with aural polyp had been made.

On Computerised Tomography scan (CT), it was a soft tissue lesion with poorly defined margins and no bony erosions noticed. Excisional biopsy of this polypoidal mass had been carried out and sent to Pathology Department for histopathological examination. On gross examination, it was a single grey white irregular soft tissue bit measuring 0.5×0.5×0.3 cm. On microscopic examination, Haematoxylin and Eosin (H&E) stained section showed tumour tissue comprising of sheets and nests of round to oval cells with atypical nuclear features e.g., large pleomorphic hyperchromatic nuclei, high nucleocytoplasmic ratio (N/C ratio) and scanty eosinophilic neoplasm [Table/Fig-1A,B]. On the basis of these features a diagnosis of round cell tumour had been made. The IHC had been applied, in which tumor was positive for Desmin [Table/Fig-1C,D] and Vimentin [Table/Fig-1E]. Tumour was negative for CK, LCA [Table/Fig-1F], chromogranin and synaptophysin. After these IHC interpretations tumour diagnosis was confirmed as RMS (Embryonal type). Patient was referred to higher centre for chemotherapy and radiotherapy.

DISCUSSION

Soderberg F, first described a case of RMS in middle ear and mastoid [1]. Approximately 30% of paediatric RMS occur in head and neck, including skull base, nasal cavity and nasopharynx [2]. However, RMS of middle ear and mastoid are rare (only 3%) [3,4]. The RMS is the third most common tumour in paediatric patients after neuroblastoma and nephroblastoma [5,6]. About 63% of RMS occurs in children below age of 10-year-old, with a peak between 2-5-year-old [7].



[Table/Fig-1]: Histological and IHC features of tumour (A) Photomicrograph shows sheets and nests of round to oval tumour cells (H&E, 100x); (B) Higher magnification shows large pleomorphic nuclei, high N/C ratio of these tumour cells (H&E, 400x); (C) Photomicrograph shows strong immunopositivity of tumour cells for desmin (IHC, 100x); (D) Desmin positivity on higher magnification (IHC, 400x); (E) Vimentin also shows strong cytoplasmic activity (IHC, 400x); (F) Photomicrograph shows negative immunoreactivity of tumour cells for LCA (IHC, 400x).

Presentation of RMS of the middle ear includes purulent or blood stained discharge, hearing loss, polypoidal mass in external auditory canal and granulation tissue. So, clinical diagnosis may be delayed because of similar clinical features as CSOM, so most of these patients are initially treated with antibiotics [8]. Menzies-Wilson R et al., presented a case of middle ear RMS in four-year-old boy, clinically diagnosed as CSOM and treated with local and systemic antibiotics [9].

According to International classification of RMS, these tumours are histologically categorised in five categories- Embryonal, Botryoid, Spindle cell, Alveolar and Undifferentiated [2]. Embryonal RMS is the most common (accounting 70-80%), while alveolar type is second most common. Embryonal type RMS have best prognosis, while alveolar type have worst prognosis. Children have better prognosis than adults [4]. Tumours originating in the middle ear are often manifested as chronic otitis media and only granulation tissue will be obtained on superficial biopsy, which may lead to delayed diagnosis. [10]. Hayes SM et al., described a case of middle ear RMS presented with right sided CSOM with a right sided Horner's syndrome [11]. Beghdad M et al., reported a case of embryonal RMS in a three-year-old child, who presented in emergency with left parotid and retroauricular swelling and facial nerve palsy [12]. Akbar A and Sohail A presented a similar case of three year old male, who presented with otalgia, blood stained discharge and polypoidal mass in right external auditory canal [5]. Bhargava S et al., also published a rare case report in adult, however with same presentation [13]. Biopsy is the mainstay of diagnosis. Biopsy should be performed as soon as possible. On histopathological examination, RMS showed features of round cell tumour [14]. In the present case, diagnosis of round cell tumour had been made and IHC had been advised to differentially diagnose the tumour from another round cell tumour (Lymphoma, Ewing's Sarcoma etc.,). Neuroendocrine markers had been applied along with vimentin, desmin and LCA. Desmin positivity of round cell tumour is sensitive and specific of RMS [8,14]. Less than one-third patients of RMS survived in 1960's; but now-a-days, rate of cured cases have improved up to 70%, largely reflecting advances made by Intergroup RMS study committee [6].

CONCLUSION(S)

The RMS of the middle ear is rare and often presents as recurrent otitis media. Diagnosis, if missed, facial nerve involvement, local meningeal and distant metastasis are common. The present case report highlights the difficulty in diagnosis of middle ear soft tissue sarcoma. The RMS in middle ear is rare, and it is associated with high mortality if diagnosed late. In the present case, patient presented with features of CSOM clinically and radiologically. However, on biopsy, diagnosis of round cell tumour had been made, which was confirmed as RMS after IHC application.

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PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of Pathology, Government Medical College, Kannauj, Uttar Pradesh, India.
2. Associate Professor, Department of Pathology, Uttar Pradesh University of Medical Sciences, Saifai, Etawah, Uttar Pradesh, India.
3. Assistant Professor, Department of Pathology, Uttar Pradesh University of Medical Sciences, Saifai, Etawah, Uttar Pradesh, India.
4. Associate Professor, Department of Pathology, Uttar Pradesh University of Medical Sciences, Saifai, Etawah, Uttar Pradesh, India.
5. Assistant Professor, Department of Pathology, Uttar Pradesh University of Medical Sciences, Saifai, Etawah, Uttar Pradesh, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Sanjeev Kumar Singh,
Associate Professor, Department of Pathology, Uttar Pradesh University of Medical Sciences, Saifai, Etawah-206130, Uttar Pradesh, India.
E-mail: kumar.sanjiv2@gmail.com

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