

## EMBRYONAL RHABDOMYOSARCOMA OF TEMPORAL BONE TWO CASE REPORTS

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### Introduction

Embryonal Rhabdomyosarcoma of the temporal bone is a rare, highly malignant tumour. It is primarily a disease of the first decade of life. Upto 1976 only 72 cases of Rhabdomyosarcomas of the ear and mastoid had been reported.<sup>1</sup> We have come across two such cases and both presented as aural polypi.

### Case Report No. 1

S.A., A five years old male child was seen in ENT O.P.D. of DHQ Teaching Hospital, Abbottabad on 25-9-1984 with six months history of blood stained discharge from right ear, right otalgia and swellings in front of and below the right pinna. Past history indicated that the patient had presented with complaints of right blood stained otorrhoea with otalgia and a mass protruding from the right ear in March, 1984. He had right aural polypectomy done in March, 1984. The swelling in the ear recurred within two months and another swelling appeared in front of the right pinna. A modified radical mastoidectomy was done in July, 1984 in DHQ Hospital, Abbottabad. On examination, the patient had right lower motor neurone complete facial paralysis. There was post-auricular scar of previous mastoidectomy. There was a single hard swelling in right pre-auricular region and another hard swelling below the right pinna which was fixed to the sternomastoid muscle. Both swellings showed no signs of acute inflammation. Right ear canal showed a polyp. The rest of E.N.T. and systemic examination was normal.

A provisional diagnosis of right CSOM with Zygomatic and Bezold's abscesses was made. The mastoid was explored the same evening. It showed the whole mastoid cavity filled with granulations and polypi, eroding sinus plate and attached to the sinus wall. The middle fossa dura was exposed. These granulations and polypi were continuous via the tip of the mastoid with the swelling below the pinna and through the ear canal with the pre-auricular swelling. Pressure on these swellings resulted in extrusion of continuous cord like masses of tissue into the mastoid cavity. The striking findings during this exploration were; very extensive granulations and polypi whose complete removal was extremely difficult and complete absence of pus both in the mastoid cavity as well as the swellings in front of and below

the right pinna. All the tissues removed were sent for histopathology which were reported as Embryonal Rhabdomyosarcoma of the temporal bone. The patient was sent for postoperative radiotherapy plus chemotherapy but did not return for follow up.

## Case Report No. II

B.A., A five years old male child presented on 7-3-85 with complaints of right blood stained ear discharge and ear ache for four months and deviation of the angle of the mouth to the left side for ten days. Past history showed nothing significant. On examination, the child was irritable, and ill-looking. He had a right aural polyp with right complete, lower motor neurone facial paralysis. The rest of the ENT and systemic examination was normal.

The right ear was explored and it showed granulations and polypi filling the mastoid and middle ear cavity. No pus was found. There was no involvement of dura or lateral sinus. The facial ridge was eroded and the VII nerve was exposed. Keeping in view the short duration of history, the extensive destruction and the large number of polypi and granulations the specimen removed was sent for histopathology, whose report showed Embryonal Rhabdomyosarcoma. The patient was sent for post-operative radiotherapy plus chemotherapy but did not come back for followup.

## Discussion

The majority of Rhabdomyosarcomas take their origin in peripheral soft tissues. In children however there is marked predilection for Rhabdomyosarcomas to present in the head and neck.<sup>2</sup> In the head and neck, Rhabdomyosarcomas occur most commonly in the orbit, with the second most common site being the soft tissues of the neck itself. Next in order of decreasing frequency are the nasopharynx, face, ear, tongue and palate.<sup>3</sup>

Histologically, Embryonal Rhabdomyosarcomas show a tendency for the tumour cells to form islands or broad cords, separated by fibrovascular stroma. The preponderant cell type is round or oval with scant cytoplasm. Scattered within this background are more characteristic strap cells, racquet-shaped cells or giant cells, having more abundant cytoplasm and myofibrils as well as cross striations.

The TNM classification of Rhabdomyosarcomas of head and neck is as follows.<sup>4</sup>

- T1 — Localized to one site.
- T2 — Extension to adjacent structures or two or more sites.
- T3 — X-Ray evidence of bone destruction or involvement of cranial nerves.
- N0 — No clinical evidence of lymph node involvement.
- N1 — Single clinically positive lymph node, less than 3 CM in diameter.
- N2 — Single lymph node more than 3 CM in diameter or multiple ipsilateral, palpable lymph nodes.

- N3 — Fixed or bilateral lymph nodes.
- M0 — No distant metastasis.
- M1 — Distant metastasis present.

Embryonal Rhabdomyosarcoma of the temporal bone is a very rare, lethal tumour. In temporal bone it was first described by Soderberg in 1932.<sup>5</sup> Since then, upto 1976 only 72 cases had been reported in world literature. It is found in children and young adults. The tumour rapidly erodes the surrounding structures and may invade the cranial cavity. These tumours possess a sufficiently rapid rate of growth that medical attention is sought within six months, usually before two months.<sup>1</sup> Clinically it presents with ear ache, blood stained ear discharge and extensive granulations and polypi in the ear. Diagnosis depends upon a high index of suspicion plus sending all suspicious material for histopathology. Treatment is combined surgery plus radiotherapy (6000 rads) plus chemotherapy, consisting of vincristine, cyclophosphamide and dactinomycin. Radical Surgery is almost impossible but surgical debulking if feasible is an important component of management because adjuvant radiotherapy, chemotherapy and immunotherapy are of maximum benefit when the body burden of tumour cells is least. The average survival time has increased over the recent past but the tumour is still uniformly fatal.<sup>6</sup>

### Summary

Two cases of Embryonal Rhabdomyosarcoma of the temporal bone are presented. Both patients were five years old and both presented with blood stained otorrhoea and otalgia, complete L.M.N. facial paralysis and aural polypi. Evolution of the disease was very rapid. Extensive bony destruction and soft tissue invasion was seen within 4-6 months of onset. On mastoidectomy, granulations and polypi were very extensive, going in every direction, with minimum suppuration. These features led to our suspicion of malignancy and that was substantiated by histopathology.

### REFERENCES

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