Malignant Schwannoma of the Sinonasal Tract

G. K. Maheshwari, Assistant Professor, Dept. of Radiation Oncology, H. A. Baboo, Professor and Head, Dept. of Radiation Oncology, Usha Gopal, Lecturer, Dept. of Radio Diagnosis, D. B. Balar, Professor and Head, Dept. of Pathology, N. M., Shah, Lecturer Dept. of Pathology
The Gujarat Cancer and Research Institute NCH Campus, Asarwa, Ahmedabad, India. Pin - 380 016

Abstract

Malignant Schwannoma of the sinonasal tract is an extremely rare tumour. So far, only 19 cases have been reported in the literature. We report herein a case of malignant schwannoma involving nasal cavity, maxillary antrum and ethmoid sinus in a 60-year-old male patient. The patient was treated with surgery and post-operative radiotherapy. He currently remains free of the disease 15 months after the diagnosis and twelve months after completing therapy.

Malignant Schwannoma originating from the sinonasal tract is an extremely rare event. The first case of malignant Schwannoma originating from maxillary antrum was reported by Millard and Busser (1952). Till date, only 19 case of malignant Schwannoma of the sinonasal tract have been reported in the literature. The origin of sinonasal malignant Schwannoma most likely is to be from the trigeminal nerve (Vth cranial nerve). It is locally aggressive tumour. Though surgery is mainstay of treatment, local recurrence is common. Adjuvant post-operative radiotherapy may help in preventing local recurrence.

In view of rarity of the condition following case of malignant Schwannoma of the sinonasal tract is being reported.

Case Report

A 60-year-old male patient presented in August 1995, with prolonged history of nasal discharge and obstruction, swelling and pain over right side of the face. On examination, the patient had his nasal septum deviated to the left side and right nasal cavity was full of secretions and a growth. His vision was unaffected and there was no cervical lymphadenopathy. There were no stigmata of neurofibromatosis (Von Recklinghausen's disease).

Plain radiography showed haziness in the right maxillary antrum, ethmoid sinus and the nasal cavity.

There was no erosion or destruction of the bony walls. CT scan of paranasal sinuses (Fig. 1 & 2) showed a very big soft tissue mass in the right maxillary antrum, nasal cavity and ethmoid sinus. There was no orbital involvement or intracranial extension of the disease on CT scan evaluation. An intranasal biopsy (Fig. 3) was reported as malignant spindle cell tumour within background of myxoid stroma. Malignant cells showed varying configuration and hyperchromatic, large nuclei. The squamous mucosa showed granulation, necrosis and severe squamous dysplasia. Immunohistochemistry revealed positive reactivity against S-100 protein and Vimentin. Final diagnosis of malignant Schwannoma (neurogenic sarcoma) was made. Initial metastatic work-up was also negative.

The patient underwent a right medial maxillectomy and ethmoidectomy with total removal of the tumour from the involved sinuses and nasal cavity. A course of post-operative radiotherapy delivering 6000 CGy in 30 fractions over a period of six weeks by tele-cobalt therapy was conducted. Twelve months post-treatment, the patient currently remains asymptomatic and there is no clinical or radiological evidence of recurrence.

Discussion

Malignant Schwannoma is an uncommon tumour, accounting for about 10% of all soft tissue sarcomas. It is also termed as malignant peripheral nerve sheath

Fig. 1 CT scan (Axial and Coronal sections) Showing soft tissue mass in right maxillary antrum, nasal cavity and ethmoid sinus.

Fig. 2 CT scan (Axial and Coronal sections) Showing soft tissue mass in right maxillary antrum, nasal cavity and ethmoid sinus.

tumour (MPNST), neurofibrosarcoma, and neurogenic sarcoma. It originates from the nerve sheath of peripheral nerves and all the cranial nerves except the optic and the olfactory nerves which lack nerve sheaths (Harkin and Reed, 1969). The most common primary sites of this tumour are extremities, retroperitoneum and trunk in decreasing order of frequency (Pizzo et al, 1993). Head and neck is an unusual site for malignant Schwannoma where it has been reported to occur in the neck, sinonasal tract, parotid region, scalp, chin, cheek, orbit etc. (Vege et al, 1994 ; Tanwar et al, 1995). It commonly occurs between third and sixth decade, but occurrence at any age is known (Ghosh et al, 1973). Significant male preponderance has been reported (Vege et al, 1994; Perzin et al, 1982)

Malignant Schwannoma originating in the sinonasal tract is an extremely rare event. Millard and Busser (1952) reported the first case of malignant Schwannoma originating from maxillary antrum in a 42-year-old female patient. Since then 18 more cases of malignant Schwannoma of sinonasal tract have been added in the literature (Perzin et al, 1982; Maroun et al, 1988; Kameyama et al, 1987; Hellquist and Lundgren 1991; Nagayama et al, 1993; Vege et al, 1994). Thus, so far only 19 cases have been reported in the past.

Malignant Schwannoma may arise de novo or as a result of malignant transformation of a pre-existing

neurofibroma (Von Recklinghausen's disease). The incidence of malignant transformation in the neurofibroma is reported in the range of 2.4 to 16.5 percent of all cases (Ductman et al, 1986; Leslie and Chenug 1987). The origin of sinonasal malignant Schwannoma most likely is to be from the trigeminal nerve (Vth cranial nerve), but the nerve of origin often can not be precisely identified. This may be related to both the small size of the nerve fibers in the paranasal sinuses and because a relatively large amount of space is available for unrestricted growth

Fig. 3 Microphotograph showing hyperchromatic spindle cells with wavy pattern within background of loose myxoid stroma. (Hematoxylineosin H-E), X400).

(Robitaille et al, 1975). In our patient there were no features of pre-existing neurofibromas any where in the body. The histopathological examination of the operated specimen also did not show any evidence of co-existing neurofibroma. Thus, it seems that malignant Schwannoma might have developed de novo in our patient.

Histopathologically densely packed hyperchromatic spindle cells with slender wavy or curled nuclei with mitotic activity and indistinct cytoplasm are suggestive of malignant Schwannoma (Woodruff et al, 1990). However, malignant Schwannomas show great histological variation. They have to be differentiated on biopsy material from tumours of salivary gland origin, fibrosarcoma, malignant fibrous histiocytoma, rhabdomyosarcoma, leiomyosarcoma, synovial sarcoma, spindle cell melanoma, and rarely osteogenic sarcoma (Ductman et al, 1988). In a difficult situation adjunctive use of immunohistochemistry and electron microscopy may help to arrive at correct diagnosis of malignant Schwannoma. On immunohistochemistry,

this tumour shows positive reactivity for S-100 protein and Leu-17 which are schwann cell markers (Wick et al, 1987; Erlandson and Woodruff, 1982).

Malignant Schwannoma of the sinonasal tract is locally aggressive tumour. It rarely metastasizes to regional lymphnodes, Distant metastases are also infrequent (Perzin et al 1982; Vege et al, 1994). Though the surgery is mainstay of the treatment, complete surgical ablation of the tumour may be difficult due to the extensions of the tumour into surrounding vital structures. Though survival ranging from few months to several years has been reported following surgery, local recurrence due to failure to control the disease with surgery alone is frequent (Millard and Busser, 1952; Perzin et al, 1982). Adjunctive post-operative radiotherapy has been recommended in order to delay or possibly prevent the local recurrence and thus improve the disease free survival (Hellquist and Lundgren, 1991; Vege et al, 1994).

References

1. Ducatman, B.S., Scheithauer, B. W., Piepgras, D. G., et al. (1986) . Malignant peripheral nerve sheath tumours. A clinicopathologic study of 20 cases. *Cancer* 57 : 2006-2021
2. Erlandson, R.A., Woodruff, J.M. (1982) . Peripheral nerve sheath tumours . an electron microscopy study of 43 cases. *Cancer* 49 : 273-287.
3. Ghosh, B.C., Ghosh, L., Huvos, A. G., et al (1973) . Malignant Schwannoma a clinicopathologic study. *Cancer* 31 : 184-190
4. Harkin, J.C., Reed, R.J. (eds) (1969) . Tumours of the peripheral nervous system. *Atlas of tumour pathology Series* 2, Fascicle 3. Washington, DC : AFIP
5. Hellquist, H.B., Lundgren, J. (1991) . Neurogenic sarcoma of the sinonasal tract. *The Journal of Laryngology and Otology* 105 : 186-190
6. Kameyama, Y., Maeda, H., Nakane, S., et al (1987) . Malignant Schwannoma of the maxilla in a patient without neurofibromatosis. *Histopathology* 11 : 1205-1210.
7. Leslie, M.D., Cheung, K.Y.P. (1987) . Malignant transformation of neurofibromas at multiple sites in a case of neurofibromatosis. *Postgraduate Medical Journal* 63 : 131-133
8. Maroun, F.B., Sadler, M., Murray, G.P., et al (1986) . Primary malignant tumours of the trigeminal nerve. *Canadian Journal of Neurological Sciences* 13 : 146-148.
9. Millard and Busser (1952) : Schwannome du maxillaire superieur d'evolution maligne. *Ann Otolaryngol Chir Cervicofac.* 69 : 706-708
10. Nagayama, I., Nishimura, T., Furukawa, M. (1993) . Malignant Schwannoma arising in a paranasal sinus. *The Journal of Laryngology and Otology* 107(2) : 146-148.
11. Perzin, K.H., Panyu, H., Wechter, S. (1982) : Nonepithelial tumours of the nasal cavity, paranasal sinuses and nasopharynx. A clinicopathologic study. XII . Schwann cell tumours (Neurilemoma, neurofibroma, malignant schwannoma). *Cancer* 50:2193-2202

12. Pizzo, P.A., Horowitz, M.E., Poplack, D. G., et al (1993) : *Solid tumours of childhood*, in Devitta Jr VT, Hellman S, Rosenberg SA, (eds) : *Cancer principles & practice of oncology*. (ed 4). Philadelphia, Lippincott, 1993, pp 1738-1791.
13. Robitaille, Y., Seemayer, T.A., Deiry, A.E (1975) . *Peripheral nerve sheath tumours involving paranasal sinuses : A case report and review of the literature*. *Cancer* 35 : 1254-1258.
14. Tanwar, R.K., Kumar, R., Malik, S., et al (1995) : *Orbital neurofibrosarcoma : A case Report*. *Indian Journal of Pathology and Microbiology* 38 : 91-94.
15. Vege, D. S., Chinoy, R.L., Ganesh, B., et al (1994) : *Malignant Peripheral Nerve Sheath Tumours of the Head and Neck : A clinicopathological study*. *Journal of Surgical oncology* 55 : 100-103.
16. Wick, M. R., Swanson, P.E. Scheithauer, B.W., et al (1987) : *Malignant peripheral nerve sheath tumour. An immunohistochemical study of 62 cases*. *American Journal of clinical pathology* 87 : 425-433.
17. Woodruff, J.M., Horten, B.C., Erlandson, R.A. (1990) : *Pathology of peripheral nerves and paragangliomas*. In Siverberg SS (ed) : *Principles and practice of surgical pathology*. New York, Churchill Livingstone. pp 1997-2017.

Conference News

The fifth Annual National Conference of the Association of Paediatric Otolaryngologists of India was held at Trivandrum from the 18th to 20th September 1998. About 300 delegates from all over India and abroad attended. There was a Live Surgical Workshop on the 18th at the SAT Hospital for Children, Medical College, Trivandrum. The operating surgeons were Dr. K. P. Morwani from Mumbai, Dr. Mohan Kameshwaran from Chennai, Dr. V. Anand from Pollachi, Tamilnadu. The second and third days proceedings were held in the convention center by the sea at the scenic world-famous Kovalam beach. There were panel discussions, orations, and free paper sessions in which leading figures from India and abroad participated. The guest faculty from abroad included Dr. Dessiderio Passali, Chairman, International Rhinology Societies, Dr Luissa Bullucci, Secretary, Italian Society of Paediatric ENT, Dr Danielle Nutti, Itali, Dr Iain Swan, U. K., Mr Jose Ramirez, U. S. A.

The topics discussed included the present status of surgical treatment of tonsillitis and adenoids, cholesteatomatous disease in children, newer concepts of sinusitis and its treatment, the early detection and management of impaired hearing in children, newer cocepts and advances in hearing aid technology including digital hearing aids, cochlear implants, the controversies regarding septal surgery in children and FESS in paediatric patients. The highlight of the social events was a memorable banquet under the starlit sky on Kovalam Beach on the 19th September. The open house session on the last day saw a lively discussion with active participation from the floor on a wide range of topics.

The national Deafness Day as suggested by the National Body was held on 11th November at the Panchayath Association Hall, Trivandrum under the joint auspices of the AOI Trivandrum Chapter and the ENT & Head and Neck Foundation Trivandrum. The function was inaugurated by the of Hon'ble Health Minister of Kerala A. C. Shanmugha Das. We examined 132 hearing impaired patients above 50 years of age and hearing aids were distributed to 40 patients. We had very good support from the Handicapped Welfare Association of the State and a number of good people. We will strongly stand with all attempts by the National Body to bring Deafness into a National Programme. Which was in our minds for a long time. The Press and TV gave a very wide coverage for the function and some of the leading Newspapers like 'The Hindu' had leading articles on the subject and its importance.

Dr. S. Madhavan Nair
Org. Secretary