Malignant Transformation in a Benign Encapsulated Schwannoma of Retropharyngeal Space : A Case Report

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Abstract

Malignant schwannomas are rare in the head and neck, however benign schwannoma of this area are common. Cases of malignant schwannoma have been reported in the nasal cavity, paranasal sinuses, eye, parapharyngeal space and neck with or without an association of von Recklinghausen disease. We described a case of an asymptomatic solitary malignant schwannoma arising in the retropharyngeal space which was treated by local excision and postoperative radiotherapy. To our knowledge, this case may be the first reported case in the world literature.

A potential retropharyngeal space is located behind the pharynx. It lies between the buccopharyngeal fascia anteriorly and the ala fascia posteriorly. It extends from the base of the skull to the level of the 6th cervical vertebra where the ala fascia fused anteriorly with its buccopharyngeal counterpart. The retropharyngeal space contains lymph nodes which drain the nose, paranasal sinus and nasopharynx(1). Bulging of the posterior pharyngeal wall, an uncommon finding, may be associated with a retropharyngeal space abscess, tuberculosis, and lymphadenopathy especially related to metastasis from cancers in the nose, paranasal sinus, or nasopharynx(2). To our knowledge, a schwannoma has not yet been reported to arise in the retropharyngeal space. We record herein a retropharyngeal benign schwannoma with focal malignant transformation. The retropharyngeal location of the current lesion is regarded as unique.

CASE REPORT

A 38-year-old Thai man was referred from a private ENT clinic for further management of a mass in his throat. The lump was accidentally found in an ENT examination for his "cold" symptoms by one of the authors (P.S.). The patient denied any oropharyngeal symptoms. Sore throat or any foreign body like sensation in his throat was not noticed

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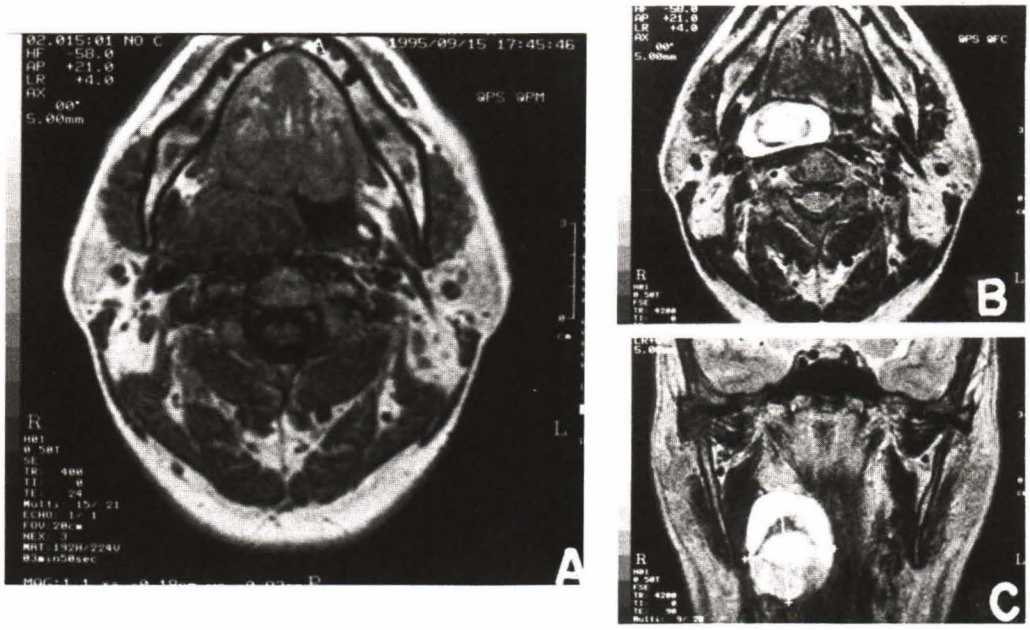

Fig. 1. A 4.10 x 5.10 x 2.5 cm well defined mixed iso/hyposignal T1 (A) and mixed hyper-hyposignal T2 (B, C) mass in the right retropharyngeal space at the oropharyngeal level.

except for a 1-week "cold" ailment which resulted in postnasal drip and frequent clearing of his throat. Neither dysphagia, odynophagia, lump in the throat nor snoring was experienced. Examination of the neck revealed injected pharyngeal mucosa with a submucosal mass bulging from the right posterolateral region of the pharyngeal wall. The mass was behind the posterior pilla of the right tonsil without either anterior displacement of tonsil or medial shifting of uvula. The exact location of the mass was noted superiorly at the upper pole of the right tonsil and inferiorly at the level of 2 cm below the lower pole of the tonsil. Laterally the mass was limited by the lateral pharyngeal wall. Medially it extended to the plane of 1 cm beyond the midline. The mass was rubbery. Bimanual palpation yielded a negative result. All cranial nerves were intact. Horner's syndrome and stigmata of von Reklinghausen's disease were absent.

MRI study showed a well-defined mass in the right retropharyngeal space at the level of the oropharynx. The lesion was nonhomogeneous and submucosally located deep to the constrictor muscle. The carotid arterial system was intact. (Fig. 1)

Surgically, the patient lay on his back in the tonsil position. The tumor was transorally

Fig. 2. Macroscopic features of schwannoma. The tumor has nodular and encapsulated external surface (upper portion). Sectioning shows homogenous grey white cut surfaces interspersed by smaller intense bright yellow foci (lower portion). The arrows point toward a necrotic zone.

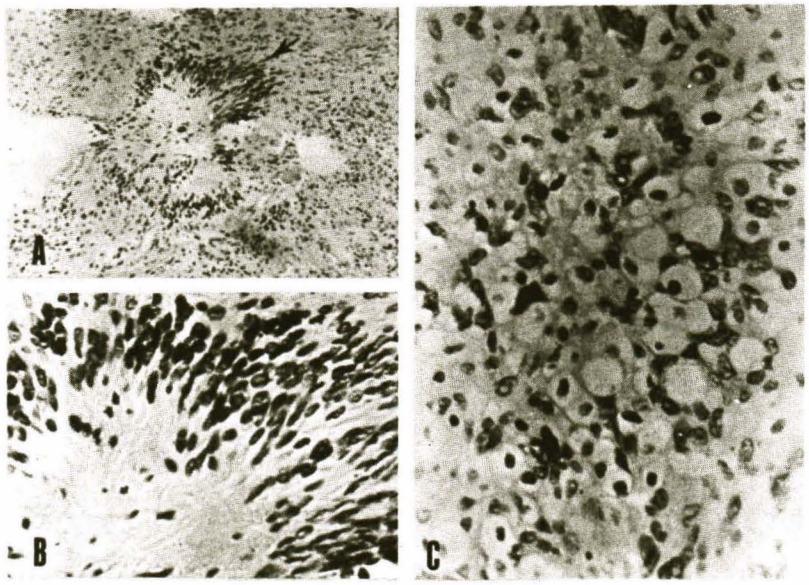

Fig. 3. Microscopic features of schwannoma, benign part.
A. Many spindle-shaped tumor cells are arranged in palisading pattern (arrow). Note a few cystic foci H & E, x 40.
B. A nuclear palisading arrangement corresponding to the arrow in A is further demonstrated. H & E, x 100.
C. Foamy histiocytes in the tumor are depicted. H & E, x 100.

excised by blunt dissection under general anesthesia. There was no postoperative complication.

Pathologic Examination

Macroscopically, the circumscribed, nodular, encapsulated, and rubbery neoplasm measured 3 x 4 x 4 cm. Its cut surfaces were smooth gray to yellow and interspersed by a few cystic changes. In an area, however, the tumor was necrotic (Fig. 2).

Microscopically, the lesion consisted of benign and malignant portions. The benign part was characterized by many spindle-shaped cells being arranged in intertwined bundles. Occasional spindle-shaped cells showed distinct nuclear palisading. Foamy histiocytes were numerous; they were in clusters, or were disseminated randomly (Fig. 3). Blood vessels with thick and hyalinized walls were noted frequently as were granules of hemosiderin.

The necrotic area indicated by the arrows in Fig. 1 was composed of many bizarre neoplastic cells with either uninucleated or multinucleated type. Bizarre hyperchromatic nuclei were also observed as were approximately 3 mitoses per 10 high-power microscopic fields (Fig. 4). Immunopathologically, both spindle-shaped cells and bizarre tumor cells showed positivity of S-100 protein in the perikaryon (Fig. 5). This part of the tumor was considered as malignant.

The pathologic impression, then, was a schwannoma with focal malignant transformation; it was considered to be similar to a few previously reported cases[3-5].

Follow-up Study

The patient further received a radical dose of radiotherapy (6000 cGy) to the tumor bed to eradicate the probable residual tumor and to prevent a chance of recurrence, and 5000 cGy to the cervical node bearing area. He tolerated the treatment well, and recovered later from the radiation effect in the next 6 weeks. He was regularly followed-up for 6 months, and there was no recurrence.

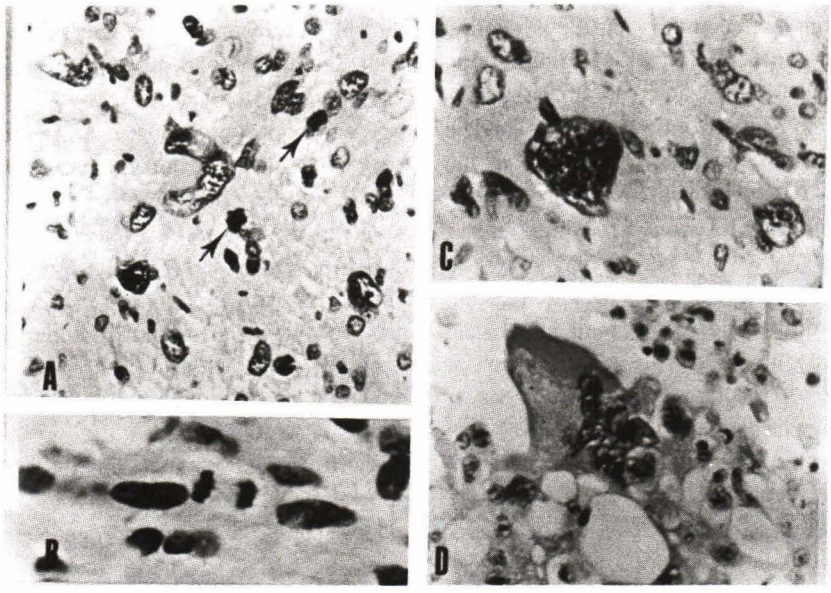

Fig. 4. Microscopic features of schwannoma, malignant part.
A. Many pleomorphic tumor cells and two mitotic figures (arrows) are shown. H & E, x 100.
B. A mitotic figure is further exhibited. H & E, x 1000.
C. and D. Considerable bizarre giant neoplastic cells are demonstrated. H & E, x 400.

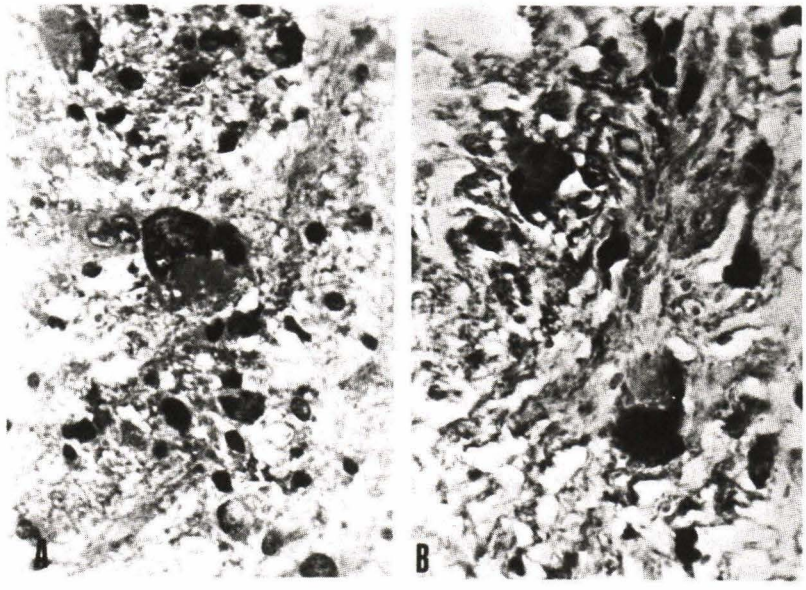

Fig. 5. Immunohistochemical features of schwannoma. A and B. Bizarre neoplastic cells with positive cytoplasmic S-100 protein represented as dark perikaryon are shown. Immunostain for S-100 protein, x 400 each.

DISCUSSION

Malignant schwannoma or the so-called nerve sheath sarcoma, malignant neurilemmoma, neurofibrosarcoma or neurogenic sarcoma is a neoplastic entity which arises from Schwann cells. The latter have been regarded as the lining cells of the nerve sheath[6]. It may arise as a solitary growth or in an association with von Recklinghausen's disease. The latter may carry a poor prognosis[7-9].

Malignant schwannomas are uncommon and account for only 5 per cent of all soft tissue sarcomas, affecting all ages with a preference for the extremities[10,11]. Only 9 to 14 per cent of them are found in the head and neck area[5-8,12]. These figures suggest no or little relation between the malignant and benign schwannomas which are more common in this region (44.8%). Most authors believe that malignancy did not transform from the benign tumor but controversies still exist[4,5, 8,13]. In the cervico-facial region, schwannomas were reported to occur in the neck, parapharyngeal space, cheek, orbit, nasal cavity, paranasal sinuses and ear. They, however, have not yet been found in the retropharyngeal space[14,15]. Our current case may be the first reported retropharyngeal schwannoma.

Identification of the nerve of origin of these tumors is usually unsuccessful, even in those cases with neurologic symptoms[6]. Frequently, the nerves adjacent to the tumor are stretched over the mass and result in neural deficits. Clinical presentation may either be a painless, gradually growing mass (73%) or a painful tumor (9%)[13]. Some patients complain of pain along the course of the peripheral nerves. The mass in our patient was surprisingly asymptomatic, eventhough it was sizable. It was accidentally noticed in the examination for a cold ailment.

The definite diagnosis of a malignant schwannoma is based on the coexistance of a tumor with a nerve fiber. Nevertheless, the nerve of origin of the tumor may be unidentifiable, even in autopsy, because of its complete destruction of the growth or due to the enormous size of the mass which prevents discovery of the nerve of origin in examination of the specimen[7]. The spindle-shaped tumor cells are often arranged in fascicles and solid cellular area. There is a moderate degree of cellular pleomorphism. Necrosis may be present. Mitotic figures are frequent[10,14]. When these typical features cannot be confirmed because of the undifferentiation and high pleomorphism, immunohistological examinations with S-100, antiglial fibrillary acidic protein, vimetin and cytokeratin should be investigated to confirm malignant schwannoma and to differentiate it from other tumors[15]. Histological grading is generally considered to be the most important factor in determining the prognosis of patients with malignant schwannoma, and is the primary factor considered in staging. Tumor grade is based upon such criteria as cellularity, pheomorphism and mitotic activity[14,16]. Aggressive wide excision which includes the involved neurovascular structures, subcutaneous tissue and underlying bone as well as adjacent uninvolved musculofascial planes results in a better prognosis compared with a less aggressive excision. Involved nerves should be resected proximal to the tumor site in an attempt to obtain clear margins. Failure to perform adequate wide excision is usually associated with an unacceptable high local recurrent rate[14,16,18].

When radical wide excision is not feasible, a local excision should be applied in conjunction with the radiotherapy. Lymph node dissection is not warranted unless metastasis is present[11]. More recently, postoperative radiation therapy has been recommended in order to decrease the incidence of local recurrences, to treat isolated recurrences, or as a primary therapy for unresectable lesions[19-21]. Furthermore, adjuvant radiotherapy may improve the 5-year survival by 20 per cent[14,16].

In our case, the tumor was in the retropharyngeal space in which a wide excision was not feasible. The tumor was totally extirpated transorally without any difficulty, and we initially believed that it was a benign tumor. Unfortunately, a schwannoma with malignant transformation was pathologically established later. The patient denied any further surgical intervention but accepted a full course of postoperative radiotherapy. He was in good health without any sign of recurrence up to the 6-month postoperative period.

In malignant schwannoma various chemotherapeutic agents have also been applied, singly or in combination, but the overall results are poor [10,22]. We decided not to give any chemotherapeutic agent to our patient because of its reported poor rate of success as well as absence of distant metastasis.

Several factors may determine the prognosis of the patient, such as the tumor size, histologic grading, nodal or distant metastasis, feasibility of aggressive excision, and adjuvant radiation therapy (+/- chemotherapy)[11,14,23]. A small tumor should be easily controlled with wide excision[23]. Generally, local recurrence for sarcomas of the head and neck varies between 17 per cent to 53 per cent after surgical removal, but repeated excision for local recurrence can improve survival and even be curative[14]. Various immunological and molecular biological approaches have been extensively studied but their uses remain investigational[14,24]. However, the long term prognosis for these neoplasms is poor regardless of various therapeutic modalities used[10].

SUMMARY

A rare case is reported of a benign encapsulated schwannoma with a focus of malignant transformation occurring in the retropharyngeal space of a 38-year-old man. The retropharyngeal location of the current schwannoma is unique. The lesion was totally removed transorally. The patient further received postoperative radiotherapy. He was in satisfactory condition after the 6-month follow-up study.

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REFERENCES

1. Hollinshed WH. Anatomy for Surgeons : The Head and Neck. 3rd ed. vol. 1. Philadelphia : Harper & Row, 1982: 269-89.
2. Ballenger JJ. Infections of the fascial spaces of the neck and floor of the mouth. In: Ballenger JJ. Diseases of the Nose, Throat, Head and Neck. 14th ed. Philadelphia : Lea & Febiger, 1991: 235-42.
3. Carstens PHB, Schrodt GRS. Malignant transformation of a benign encapsulated neurilemmoma. Am J Clin Pathol 1969; 51: 144-9.
4. Hanada M, Tanaka T, Kanayama S, et al. Malignant transformation of intrathoracic ancient neurilemmoma in a patient without von Recklinghausen's disease. Acta Pathol Jpn 1982; 32: 527-36.
5. Yousem SA, Colby TV, Urich H. Malignant epithelioid schwannoma arising in a benign schwannoma : A case report. Cancer 1985; 55: 2799-803.
6. Cutchavaree A, Shuangshoti S, Kumut N. Parapharyngeal neurogenic tumors : Nine-year experience. J Med Assoc Thai 1984; 67: 351-5.
7. Cutchavaree A, Shuangshoti S, Wisuthiseriwong W, Samranvej P. Sarcoma of nerve sheath of head and neck. J Med Assoc Thai 1984; 67: 448-55.
8. Shapiro MJ, Rickert RR. Malignant parapharyngeal schwannoma (neurilemmoma). Otolaryngol Head Neck Surg 1979; 87: 653-8.
9. Karmody CS. Malignant schwannoma of the trigeminal nerve. Otolaryngol Head Neck Surg 1979; 87: 594-8.
10. Elias MM, Balm AJM, Peterse JL, et al. Malignant schwannoma of the parapharyngeal space in von Recklinghausen's disease : A case report and review of the literature. J Laryngol Otol 1993; 107: 848-52.
11. Bailet JW, Abemayor E, Andrews JC, et al. Malignant nerve sheath tumors of the head and neck : A combined experience from two university hospitals. Laryngoscope 1991; 101: 1044-9
12. Bornes L. Surgical pathology of the head and neck. vol 1. New York: Marcel Dekker, 1995: 667-70.
13. Das Gupta TK, Brasfield RD. Solitary malignant schwannoma. Ann Surg 1970; 171: 419-28.
14. Greager JA, Reichard KW, Campana JP, et al. Malignant schwannoma of the head and neck. Am J Surg 1992; 163: 440-2.
15. Nagayama I, Hishimura T, Furukawa M. Malignant schwannoma arising in a paranasal sinus. J Laryngol Otol 1993; 107: 146-8.
16. Greager JA, Das Gupta TK. Adult head and neck soft tissue sarcoma. Otolaryngol Clin North Am 1986; 19: 565-72.
17. Ghosh BC, Ghosh L, Huvos AG, et al. Malignant schwannoma : A clinicopathologic study. Cancer 1973; 31: 184-90.
18. Wilson JA, McLaren K, McIntyre MA, et al. Nerve sheath tumors of the head and neck. Ear Nose Throat J 1988; 67: 103-10.
19. Hoffmann DF, Everts EC, Smith JD, et al. Malignant nerve sheath tumors of the head and neck. Otolaryngol Head Neck Surg 1988; 99: 309-14.

20. Goefert H, Lindberg RD, Sinkovics JG, et al. Soft tissue sarcoma of the head and neck after puberty : Treatment by surgery and postoperative radiation therapy. Arch Otolaryngol 1977; 103: 365-8.
21. Basso-Ricci S. Therapy of malignant schwannomas : Usefullness of an integrated radiologic surgical therapy. J Neuro Surg Sci 1989; 33: 253-7.
22. Storm F, Eilber F, Mira J, et al. Neurofibrosarcoma. Cancer 1980; 45: 126-9.
23. Ducatman BS, Scheithauer BW, Piepgras DG. Malignant peripheral nerve sheath tumors : A clinicopathologic study of 120 cases. Cancer 1986; 57: 2006-21.
24. Buchberg AM, Cleaveland LS, Jenkins NA, et al. Sequence homology shared by neurofibromatosis type-1 gene and IRA-1 and IRA-2 negative regulator of the RAS cyclic AMP pathway. Nature 1990; 347: 291-4.

มะเร็งของปลอกหุ้มเส้นประสาท : รายงานผู้ป่วย

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ผู้รายงานได้นำเสนอผู้ป่วยที่เป็นเนื้องอกชนิดไม่ร้ายแรงของปลอกหุ้มเส้นประสาท (benign schwannoma) ซึ่งมีจุดเปลี่ยนแปลงไปเป็นมะเร็งของปลอกหุ้มเส้นประสาท (malignant schwannoma) เกิดขึ้นเดี่ยว ๆ ในช่องหลังฟาริงซ์ แม้ว่าจะเคยมีรายงานว่าเกิดมะเร็งชนิดนี้ในโพรงไซนัส เบ้าตา ช่องข้างฟาริงซ์ และคอ เท่าที่ทราบยังไม่เคยพบมะเร็งชนิดนี้ในช่องหลังฟาริงซ์ ผู้ป่วยได้รับการรักษาโดยการผ่าตัดเอาก้อนเนื้องอกออก และตามด้วยรังสีรักษา จากการติดตามการรักษา 6 เดือน ไม่พบการกลับเป็นซ้ำของโรค

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