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SCHWANNOMA OF THE SUBLINGUAL GLAND : REPORT OF A CASE

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CONTENT

Schwannoma, also referred to as neurilemmoma, is a solitary, benign and slow growing tumor of nerve sheath origin. This rare lesion originates from Schwann cell of peripheral, autonomic and cranial nerve. Extracranial neurogenic tumor of the head and neck is uncommon. Schwannoma of the salivary gland is a particularly rare form of an extracranial neurogenic tumor, with most presenting in the parotid gland originating from a peripheral branch of the facial nerve. In this report, an unusual case of schwannoma in the sublingual region is presented and the literature concerning this subject is reviewed.

Ⅰ. CASE REPORT

Clinical Findings

A 20-year-old woman presented with a swelling in the left submandibular area of nine months' duration. There was no associated pain or inflammation. There was no history of past episodes of sialolithiasis and pain. Past medical and surgical histories were noncontributory. The result of the physical examination was entirely normal, apart from a firm mass sized 3 x 4 cm in the left submandibular area. The mass was neither tender nor attached to deep or overlying tissues. There was no evidence of the neurologic deficit and the associated lymphadenopathy.

Laboratory Findings

Laboratory evaluation including a complete blood counts, chemistry survey profile, prothrombin time, partial thromboplastin time and urinalysis were all normal.

Diagnostic Imaging Findings

Conventional radiographs including periapical radiographs, occlusal radiograph and panoramic radiograph showed no evidence of the bony change related to the firm mass.

Sialogram of the left submandibular gland showed the upward displacement and dilatation of the distal third of the main duct(Fig.1).

Ultrasonograph demonstrated a mass with heterogeneous and low echogeneity in the left submandibular region.

MRI showed a well circumscribed, nonhomogeneous mass sized 3 x 5cm in the sublingual region. The mass was generally isointense compared to muscle on T1-weighted images(Fig.2), but demonstrated hyperintensity on T2-weighted images(Fig.3). MRI showed the mass to be separate from the adjacent submandibular salivary gland. These MRI features gave an impression of pleomorphic adenoma, probably arised from the sublingual gland.

Macroscopic Findings

The firm mass was easily removed. The cut surface showed homogeneity in texture and well-encapsulated margin.

Microscopic Findings

Sections of the neoplasm showed an encapsulated spindle cell tumor, in which two tissue types were clearly evident. Antoni type A areas predominated(Fig.4). These consisted of densely aggregated cells. Their nuclei were aligned in sinuous rows and separated column from column by eosinophilic fibrillary cytoplasm. Verocay bodies were also present. More loosely organized, less cellular Antoni type B areas characterized by widely separated polymorphic tumor cells embedded on a myxomatous pale staining matrix were identified. Mitoses were absent. Immunocytochemical staining with anti-S-100 protein antibodies was strongly positive within the tumor cells. A diagnosis of benign neurilemmoma was rendered without difficulty.

The patient recovered uneventfully and no neurologic dificits were noted postoperatively.

Ⅱ. DISCUSSION

The schwannoma or neurilemmoma is a benign neoplasm with no known cause or stimulus[1]. About 25% of schwannomas reported have occurred in the head and neck region[2]. It is derived from the proliferation of Schwann cell(neural crest origin) of the neurilemmoma that surrounds peripheral nerves. Schwann cell is thought of as the precursor cell of the neurofibroma, the schwannoma, and most likely the neurogenic sarcoma[3]. The schwannoma is usually slow growing but may undergo a sudden increase in size, thought in some cases to be due to intralesional hemorrhage[1]. As the lesion grows, the nerve is pushed aside and does not become enmeshed within the tumor. Another important clinical feature is the extremely low rate of malignant transformation associated with the schwannoma as compared with the relatively high rate associated with the neurofibroma in neurofibromatosis[1]. So the pathologist should attempt to differentiate between these tumors.

Schwannoma has a capsule, neurofibroma generally does not. The myxoid changes as well as the highly cellular areas found in schwannoma may occur also in neurofibroma although nuclear palisading usually is not a prominent feature in neurofibroma[4]. However, of considerable clinical significance is the fact that solitary schwannoma is not seen in the syndrome of neurofibromatosis. Malignant schwannoma associated with neurofibromatosis is reported to vary from 26% to 70%[5].

Malignant schwannoma is a highly aggressive nerve sheath tumor capable of arising de novo of from preexisting benign neurofibroma. Malignant schwannoma comprises approximately 5% of all soft tissue sarcomas.

Only 8% to 14% of these tumors arise in the head and neck[6,7]. Malignant schwannoma is

not thought to arise in pre-existing benign schwannoma[8]. Patients with neurofibromatosis tend to have larger, higher grade malignant tumors, which are more likely to arise in a preexisting neurofibroma. These malignant schwannoma recurs and metastasizes more frequently when compared to sporadic solitary malignant schwannoma[9]. On MR imaging, benign schwannoma typically has smooth, well-defined margin and is ovoid or fusiform in shape[1]. These tumor is bright on T2-weighted images and demonstrate intratumoral nonhomogeneity[11]. MRI is useful in defining the extent of disease, the relationship to neurovascular structure and surrounding soft tissue and in assisting in surgical planning[10]. The microscopic features of schwannoma are usually highly characteristic with 2 different cell patterns. Antoni A pattern consist of palisading spindle-shaped Schwann cells around an acellular central region. This cluster of cells is called a Verocay body, which is characteristic of a schwannoma[12]. The fine structure of Verocay bodies has been reported to have parallel, but separte, long cell processes, converging from opposing sides to a central area containing thickened external lamina and collagen fibrils with nuclei aligned on both sides of this central area[13]. In contrast, the Antoni B pattern is less cellular, loose, lacking arrangement in bundles and palisades.

It forms no distinctive pattern. Degenerative changes, such as cystic degeneration or hemorrhagic necrosis can be present. Isolated schwannoma of the major salivary glands is extremely rare. Most of the cases reported have involved the facial nerve in the parotid gland. We were able to find 2 previously located in the submandibular salivary gland. However, in our case, surgical exploration showed the mass to be separate from the adjacent submandibular salivary gland. The hypoglossal and lingual nerves were identified and did not appear to have any direct attachment to the tumor. Both were preserved without difficulty. The schwannomas located within the sublingual salivary gland, which do not involve surrounding structures, may arise from an autonomic nerve that serves the gland, but if the nerve's origin is small it may never be identified[14]. The treatment of benign schwannoma involves complete local excision and recurrence is unlikely. Prognosis is excellent.

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-국문 초록-

설하선에 발생된 신경초종

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신경초종은 신경초에서 발생되는 성장이 느린 양성 종양으로 단발성이며 드물다. 이 종양은 말초 신경, 자율 신경, 뇌신경의 Schwann세포에서 발생된다. 두경부 영역의 두개외 신경성 종양은 드물며 특히 타액선에서 발생되는 예는 드물다.

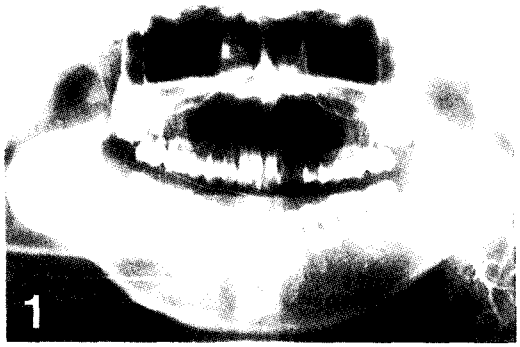
저자들은 20세 여자 환자에서 아래와 같은 소견을 나타내는 신경초종을 경험하였기에 보고하는 바이다.

1. 주소는 좌측 악하선 부위의 종창이었으며 촉진시 3 x 4 cm의 단단한 종괴가 만져졌으나 동통이나 림프절병증은 없었다.
2. 일반방사선사진에서 종괴와 관련된 골변화는 없었으나 초음파 영상에서는 불균질한 반사 양상을 보이는 종괴가 좌측 악하선 영역에서 관찰되었다.
3. 자기공명영상으로 병소의 정확한 위치를 확인할 수 있었다. 병소의 경계는 명확하였으나 신호 강도는 균일하지 않았으며 T1 강조 영상에서는 근육과 유사한 신호 강도를 보였으나 T2 강조 영상에서는 근육보다 높은 신호 강도를 나타냈다.
4. 조직병리학적으로 Antoni type B 부위보다는 A 부위가 지배적이었으며 Verocay body도 나타났다. 종양 세포는 anti-S-100 염색에 양성으로 반응하였다.

사진 부도

Fig. 1. Sialogram of the left submandibular gland showed the superior displacement and dilatation of the main duct.
Fig. 2. T1-weighted MRI image showed the isointensity of the lesion.
Fig. 3. T2-weighted MRI image showed the hyperintensity of the lesion.
Fig. 4. Histopathologic finding of the lesion showed the predominant Antoni type A areas.

논문사진부도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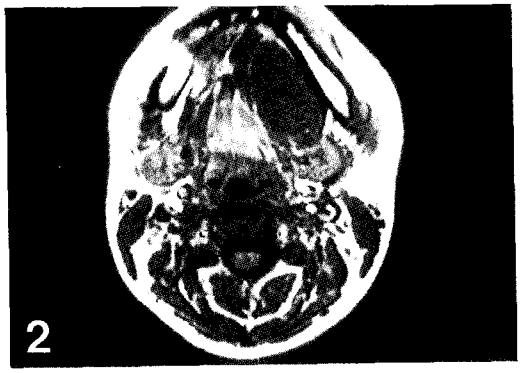
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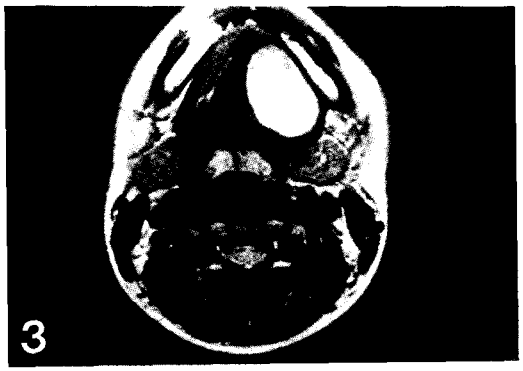
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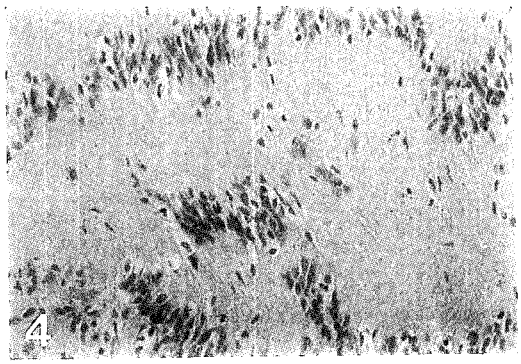
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