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Case Reports

A Case of Neurofibroma of the Cervical Region

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Neurofibroma is a benign tumor originating in the peripheral nerve sheath that may occur as part of von Recklinghausen's disease. Furthermore, multiple lesions are seen in this disease. This report describes a 25-year-old female who consulted our hospital complaining of cervical swelling, dysphagia and nocturnal dyspnea. The patient complained of a mass, which had been in that location for many years, had been causing dysphagia and nocturnal dyspnea for two or three months. The right neck and retromandibular region were swollen and elastic hard. Multiple café-au-lait spots and diffuse small pedunculated nodules were also found over the whole body. Surgical resection of the tumor was performed under a clinical diagnosis of neurofibroma associated with von Recklinghausen's disease.

Key words: Neurofibroma, Von Recklinghausen's disease, Surgical resection, Jugular foramen syndrome

INTRODUCTION

Neurofibroma accounts for nearly half of all benign nerve sheath tumors found in the head and neck region[1)]. Plexiform neurofibroma is a poorly circumscribed, diffuse enlargement of neural sheets that typically involves major nerve trunks of the head and neck region because of the rich innervation of this area[2)]. This report describes a patient with neurofibroma in the cervical region and the surgical techniques to approach the upper parapharyngeal region are discussed.

CASE

On December 4, 2001, a 25-year-old woman consulted our hospital with a chief complaint of a cervical swelling that had continuously increased over many years. The patient complained of dysphasia and nocturnal dyspnea during the recent few months. The swelling extended from the right neck to the retromandibular region and a movable elastic hard mass was palpable beneath the normal skin. Multiple café-au-lait spots and diffuse small pedunculated nodules were also found over the whole body (Fig. 1 A, B).

Incision biopsy was performed and the specimens were

Fig. 1A: Swelling in the right cervical area.

Fig. 1B: Multiple café-au-lait spots and diffuse plexiform neurofibromatosis were also noted in the lumbar region.

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submitted for histopathologic examination. The microscopic appearance of the specimen showed spindle-shaped cells intermingled with a fibrillar stroma. According to these clinical symptoms and histopathological findings, the diagnosis was neurofibroma due to von Recklinghausen's disease.

CT scans showing a well-circumscribed mass with soft tissue density, measuring 85 × 50 × 30mm. Anterior displacement of the carotid artery and internal jugular vein (Fig. 2), and stenosis of the hypopharyngeal cavity were observed (Fig. 3).

MRI showed that the top of the tumor extended into the jugular foramen (Fig. 4). On angiographic findings, the tumor did not contain any branch of the right carotid artery (Fig. 5A, B).

On March 4, 2002, surgical resection of the tumor was performed under the clinical diagnosis of neurofibroma associated with von Recklinghausen's disease. Cervical incision was used to approach the tumor (Fig. 6). The entire mass was exposed after the skin flap was raised and retracted anteriorly. The posterior belly digastric muscle was removed from the hyoid bone. The sternocleidomastoid muscle was found at the posterior edge of the tumor mass, and internal jugular vein, carotid artery and vagus nerve ran through the superficial surface of the tumor mass (Fig. 7A). The tumor mass was not adherent to the surrounding tissue and could be remove easily (Fig. 7B). Internal jugular vein, carotid artery, accessory nerve, hypoglossal nerve and vagus nerve were preserved. The histopathological diagnosis was neurofibroma based on a section from the tumor showing bundles of spindle-shaped cells within a delicate stroma without signs of malignancy (Fig. 8).

Stenosis of the hypopharyngeal cavity improved postoperatively (Fig. 9), but jugular foramen syndrome appeared

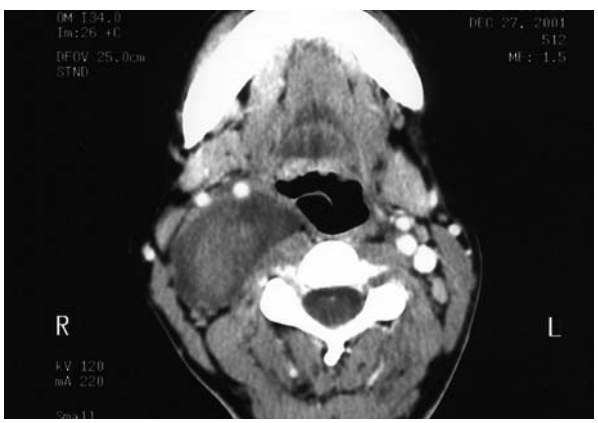

Fig. 2: CT findings. CT scans showing a well-circumscribed mass with soft tissue density.
Anterior displacement of the carotid artery and internal jugular vein was noted.

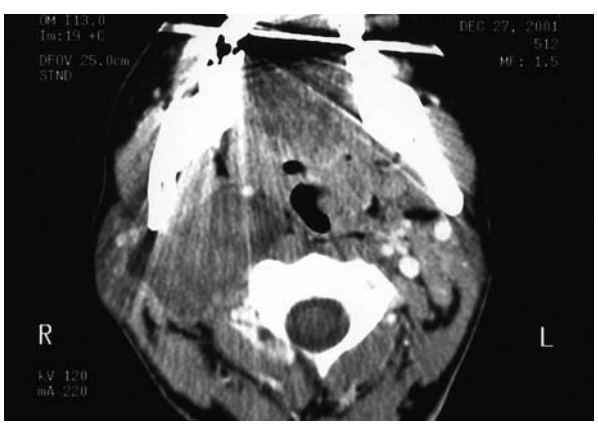

Fig. 3: CT findings. Stenosis of the hypopharyngeal cavity was observed.

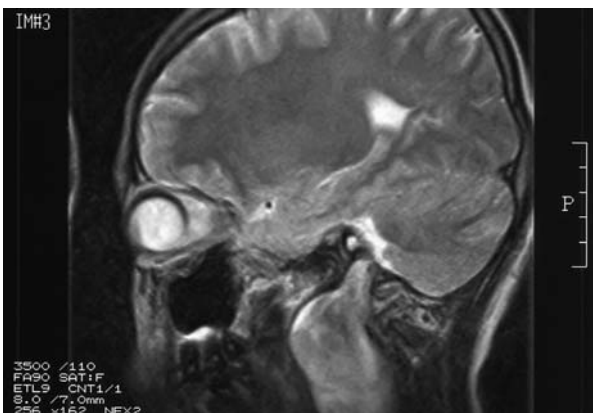

Fig. 4: MRI findings. The top of the tumor grew into the jugular foramen.

Fig. 5A, B: Angiographic findings. The tumor did not contain any branch of the right carotid artery.

Fig. 6: Diagram of the incision for direct access to the entire lesion.

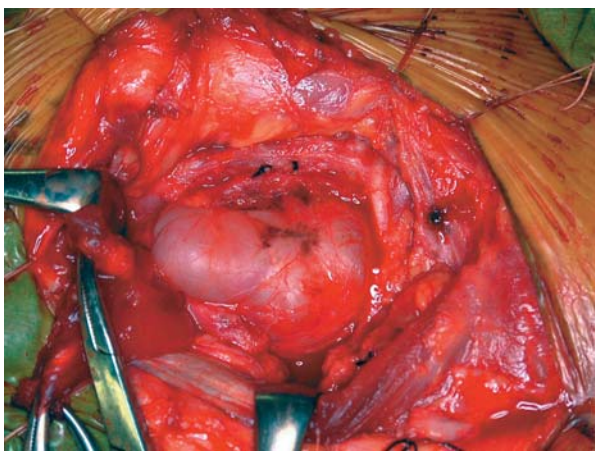

Fig. 7A, B: Excision the tumor tissue. The surgical specimen measured 85×50×30mm.

as a postoperative complication (Fig. 10). Many neurological manifestations such as hoarseness, dysphasia, misswallowing, nausea and vomiting were observed. Theses neurological manifestation improved within 2 to 3 months postoperatively (Fig. 11).

DISCUSSION

Neurofibroma is one of the most common benign tumors

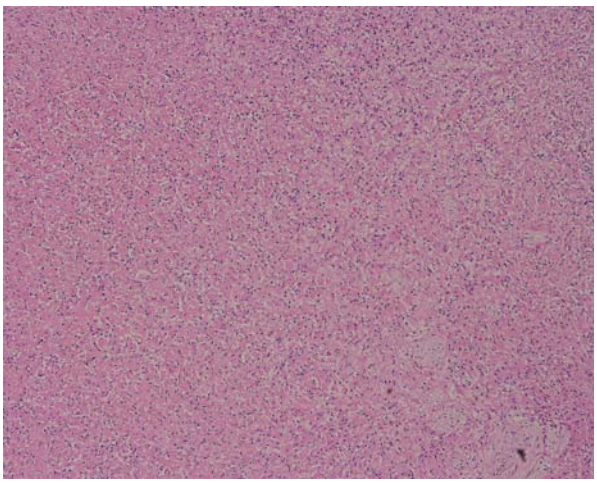

Fig. 8: Histopathological examination of the entire specimen showed that the tumor consisted of spindle-shaped cells with a fine collagenous stroma. There was no nuclear atypia.

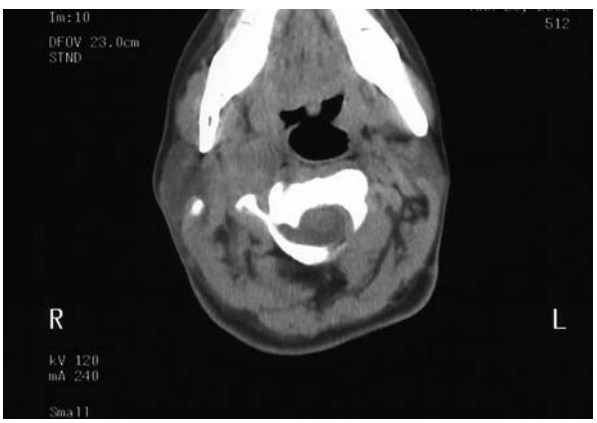

Fig. 9: Postoperative CT findings. Stenosis of the hypopharyngeal cavity was improved.

Fig. 10: Postoperatively, curtain sign of the soft palate was demonstrated immediately.

Fig. 11: Three months postoperatively.

that arise along a nerve or nervous tissue. These lesions may occur in peripheral nerve, soft tissue, skin or bone, and are derived from an admixture of Schwann cells and perineural fibroblast proliferation. Multiple lesions occur in patients with von Recklinghausen's disease (neurofibromatosis type I). Neurofibroma is a diffuse enlargement of neural sheets that typically involves major nerve trunks of the head and neck region because of the rich innervation in this area[2~4], but in the oral cavity neurofibroma generally forms a small, well circumscribed, but not encapsulated mass[5~7].

Neurofibromas are commonly treated by surgical removal[8]. Dermal neurofibromas are not usually removed unless they are painful or disfiguring, because there are generally so many of them and the lesions are not dangerous. Removal of neurofibromas can be more difficult because they can be larger and cross tissue boundaries. However, besides pain, plexiform neurofibromas are sometimes removed due to the possibility of malignant transformation[9].

An example of these can be found in the case of isolated giant plexiform neurofibroma involving all branches of the common peroneal nerve, which discusses the removal of a large plexiform neurofibroma. Sometimes plexiform neurofibromas form in locations that make the lesion especially hard to access. Especially, surgical procedures in the upper part of the parapharyngeal space, including the nasopharynx and skull base, is relatively difficult to approach because of the presence of the carotid artery, internal jugular vein and cranial nerves. Many surgical techniques to approach the upper parapharyngeal space have been reported as follows; Cervical Approach, Transparotid Approach, Cervical-parotid Approach, Transoral Approach, Infratemporal Fossa Approach, Cervical Transpharyngeal Approach, Mandibular Osteotomy, mandibular swing approach and Transmandibular Transpterygoid approach.

The lesion in this case was located in the upper parapharyngeal space, and the surgical technique chosen was the Cervical Approach because the sternocleidomastoid muscle was found at the posterior edge of tumor mass, and internal jugular vein, carotid artery and vagus nerve run through the superficial surface of the tumor mass. Using this approach, the tumor mass was removed easily and safely.

Stenosis of the hypopharyngeal cavity was improved after surgery but many neurological manifestations involving the glossopharyngeal nerve, vagus nerve, accessory nerve and hypoglossal nerve such as hoarseness, dysphasia, misswallowing, nausea and vomiting were observed (jugular foramen syndrome). However, this syndrome improved within 2 to 3 months postoperatively. For 3 years postoperatively to date, the lesion has not recurred locally.

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頸部に発生した神経線維腫の1例

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神経線維腫は頭頸部領域における良性腫瘍のうち，最もよく見られる腫瘍の一つであり，また von Recklinghausen 氏病の一病変としても発生する．Von Recklinghausen 氏病は，多発性線維腫，皮膚のカフェオレ斑および中枢神経系の腫瘍を主徴候とする疾患である．今回われわれは，この von Recklinghausen 病の部分症状として発生した，上頸部から頭蓋底にいたる巨大な神経線維腫の1例を経験したので報告する．患者は25歳の女性．右側顎下部から下顎後部にかけての腫脹および圧痛と気道閉塞感を主訴に来院した．初診時，同部に50×30mm 大の弾性硬，非可動性，境界不明瞭な腫瘤を触知した．表面皮膚は正常であるが，腫瘤のやや下方にカフェオレ斑を認めた．CT および MRI 検査で，舌骨付近から副咽頭間隙，頭蓋底にいたる80×50×30mm の境界明瞭で，T2強調像で高信号を示す腫瘍を認めた．そして腫瘍により気道が圧迫狭窄され，また頸動静脈がともに前方に圧排されているのを認めた．既往歴として，当院整形外科にて von Recklinghausen 病の診断を受け，手指および下肢の神経線維腫摘出術を施行されていた．神経線維腫の臨床診断下に全身麻酔下で腫瘍摘出術を施行，総頸動脈，内頸静脈を腫瘍より遊離し迷走神経を確認，温存しながら中枢側へ追随，下顎後部で腫瘍は顎二腹筋後腹と舌下神経の間に位置していたため顎二腹筋後腹を切離，内頸動静脈，迷走神経および舌下神経を温存しつつ，腫瘍を鈍的に剥離摘出した．術後より誤嚥や嗄声などの頸静脈孔症候群の症状が出現したが，術後およそ3ヶ月経過した時点でいずれも改善した．

キーワード：神経線維腫，レックリングハウゼン氏病，切除，頸静脈孔症候群

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