Myxoma of the Nasal Cavity and Paranasal Sinuses : Report of a Case

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Abstract

Myxomas are benign connective tissue tumors composed of stellate and spindle – shaped cells with benign – appearing nuclei lying in a myxoid stroma. They are usually found in the heart, soft tissues, and bones. Myxomas of the nasal cavity and paranasal sinuses are very rare. They are benign but of a locally invasive nature. Recurrent rate is high because of the difficulty to excise them completely. This article presents a case of myxoma of the nasal cavity and paranasal sinuses that extensively invaded the brain and orbits causing blindness. After three resections, also with a transbasal craniotomy in the second operation, the patient remains free from the disease.

Key word : Myxomas, Nasal Cavity, Paranasal Sinuses

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Myxomas are benign mesenchymal tumors that occur rarely in the nasal cavity and paranasal sinuses. They may invade locally to the bone of the skull base, orbits and brain. The recurrent rate is high because of the difficulty to resect them completely. This article presents a case of myxoma of the nasal cavity and paranasal sinuses that extensively invade the brain and orbits causing blindness.

CASE REPORT

A 23-year-old man was treated in this hospital with a 2-year history of nasal stuffiness and visual loss.

Two years previously, he complained of 3 days of sudden right visual loss, fever, proptosis of the right eye, and mucopurulent discharge from the

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nose. He was diagnosed as acute pansinusitis with orbital cellulitis. CT scan showed pansinusitis with extensive pus collection in the sphenoid, ethmoid, maxillary sinuses as well as in the nasal cavity. Compression of the right temporal lobe and the posterior aspect of the right orbit were noted (Fig. 1). The visual acuity test revealed hand movement of the right eye. Endoscopic sinus surgery under local anesthesia was then performed. Pus and tumor mass were found in the posterior nasal cavity and bilateral paranasal sinuses. Operation was terminated and the tumor mass was partially resected. Histologic examination showed myxoma with invasion to surrounding tissue (Fig. 2 and 3). He was reoperated on *via* a right lateral rhinotomy and bilateral Caldwell-Luc operation. The tumor was removed from the posterior nasal cavity, bilateral maxillary, ethmoid and sphenoid sinuses, and nasopharynx. Histologic examination confirmed myxoma with invasion to soft tissue and bone.

He was discharged from the hospital 10 days later. During the follow-up period, he was in

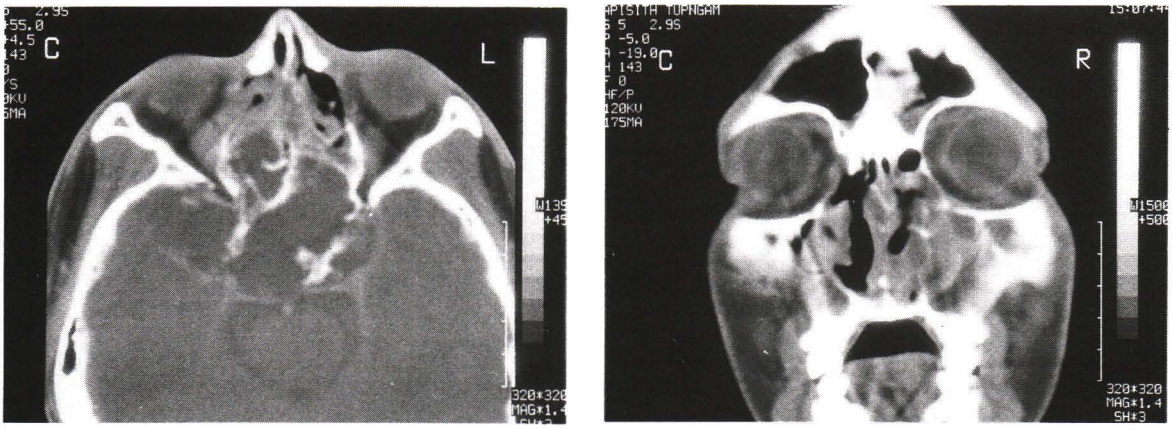


Fig. 1. CT scan at the first admission shows right pansinusitis and compression of the right temporal lobe and orbit.

Fig. 2. Histologic examination reveals tumor cells composed of spindle and stellate cells dispersed in myxoid stroma. Invasion to surrounding soft tissue is noted. (PAS stain, original magnification, x 25)

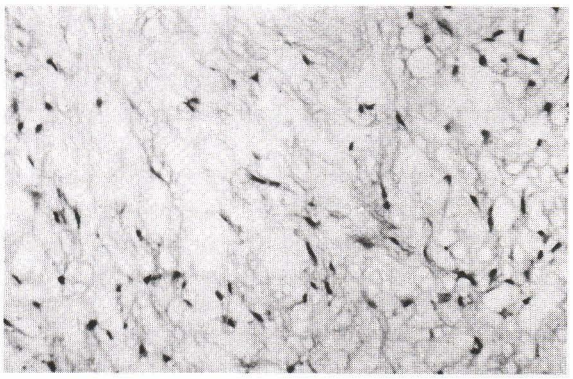
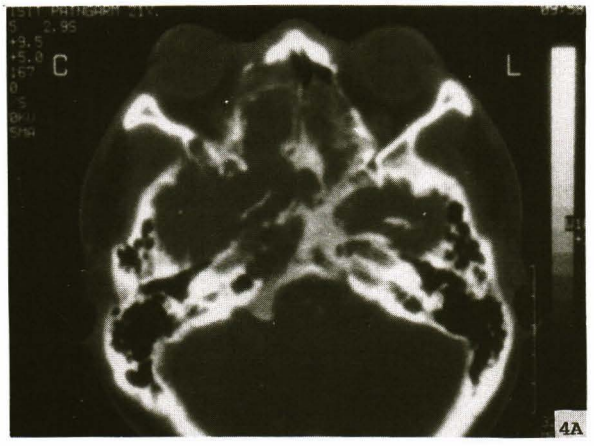


Fig. 3. Tumor cells show stellate shape lie in myxoid ground substance. (Mucicarmine stain, original magnification, x 160)



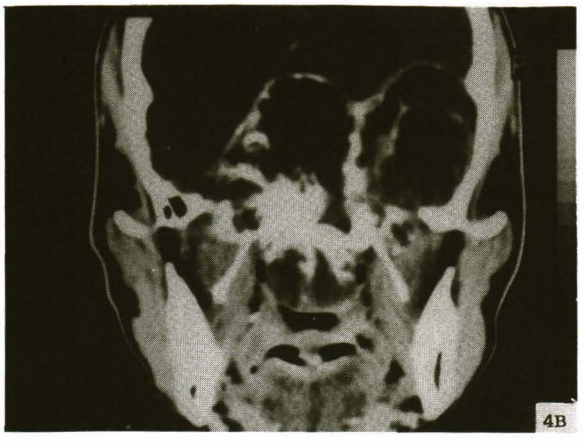
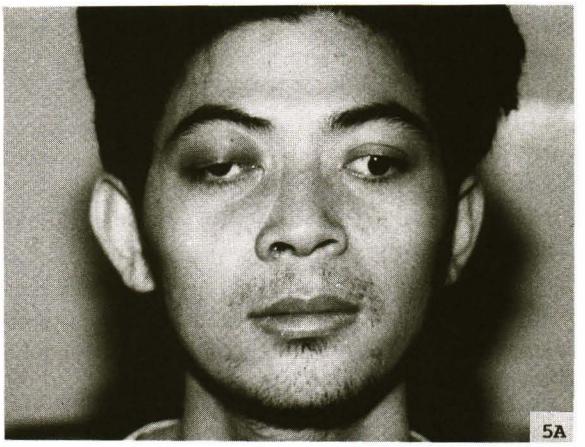
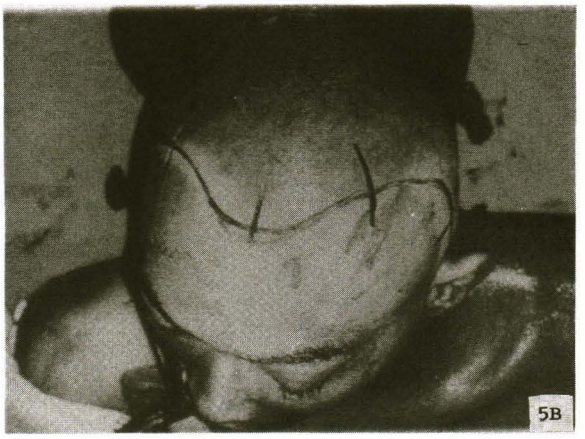


Fig. 4. CT scan shows the mass extensively involving the paranasal sinuses, posterior nasal cavity, nasopharynx, and intracranial extension.

good health but the visual acuity of the right eye did not improve.

After loss from follow-up, he was readmitted 1.5 years later for recurrent nasal stuffiness of 4-month's duration and left blindness of 3-day's duration. Eye examination revealed no light perception and optic disc atrophy of both eyes. CT scan showed the recurrent mass involving cephaladly to the sphenoid sinus, suprasellar region, frontal lobe; caudally to the nasopharynx and oropharynx; anteriorly to the nasal cavity, maxillary sinus, ethmoid sinus and right laterally to the right temporal lobe. Multiple bony destructions were seen and right exophthalmos was noted (Fig. 4). The operation consisted of a transbasal craniotomy (Fig. 5) and





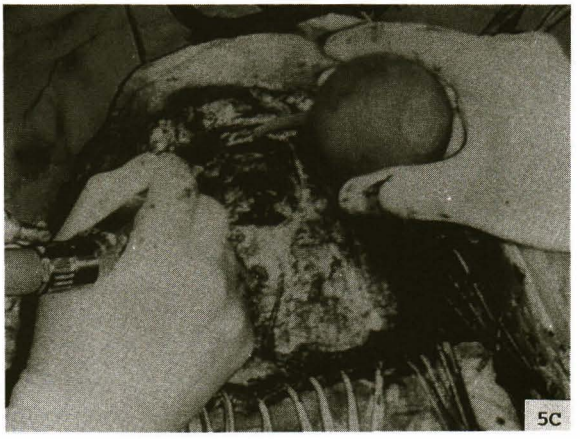


Fig. 5. Transbasal craniotomy A) Preoperative picture, B) Skin marking on the scalp, C) Tumor was removed from the right frontal lobe and temporal lobe dura.

then followed by a right lateral rhinotomy 3 weeks later (Fig. 6). The tumor filled the posterior nasal cavity, sphenoid sinus, ehtmoid sinus, right maxillary sinus, right orbital apex and adhered to the frontal lobe and right temporal lobe dura. The tumor was removed and bone graft was placed at the supra sellar region. Histologic examination still confirmed myxoma. During 15 months of the follow-up period, he was in good condition except for permanent blindness.

DISCUSSION

Myxomas are benign connective tissue or mesenchymal tumors composed of stellate and spindle - shaped cells with benign - appearing nuclei lying in a mucoid or myxoid ground substance. The degree of cellularity and amount of fibrous tissue stroma present are variable. Grossly, these tumors appear gray - white, tan or yellow and often glisten on the cut surface. They may be soft and gelatinous or firm in consistency. The tumors may appear to be encapsulated, but microscopically lack a true capsule and infiltrate irregularly into surrounding bone and soft tissue.

The etiology and histogenesis are unclear. Some authors believe that these tumors derive from primitive mesenchyme[1] while others believe that facial bone myxomas arise from dental anlage tissue [2-4]. Myxomas of the nasal cavity and paranasal simuses appear to arise from the bone.

Although myxomas are not uncommon, they are rare in the head and neck and usually involve the jaws. Stout[1] reviewed 143 cases of extracardiac myxomas, 22 of which occurred in the head and neck. Only five of these arose in the sinuses. Canalis et al[5] reported 25 cases of myxomas treated at UCLA center for the Health Sciences during a 20-year period, 4 of which occurred in the nasal cavity and paranasal sinuses. Fu and Perzin[6] reported 6 cases of myxomas from 256 non-epithelial tumors of the nasal cavity, paranasal sinuses and nasopharynx. Myxomas must be differentiated from other benign and malignant tumors that may exhibit a myxoid stroma. These tumors include Schwann cell tumors (neurofibromas, Schwannomas), fibroma, fibrous dysplasia, ossifying fibroma, liposarcoma, fibrosarcoma, rhabdomyosarcoma, chondrosarcoma, and neurogenic sarcoma.

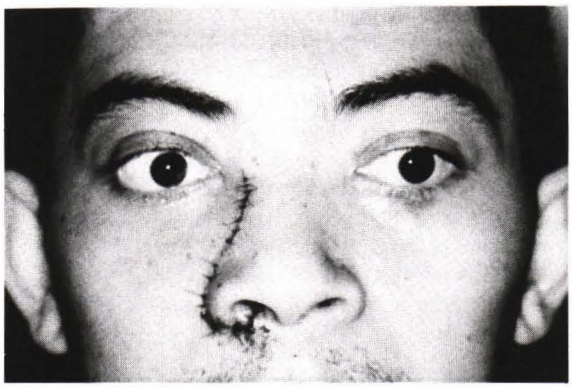


Fig. 6. Post-operative right lateral rhinotomy (Improvement of the exophthalmos was noted.)

Because of their infiltrative nature, myxomas must be treated with wide, complete resection or otherwise recurrent tumors will develop subsequently. Kountakis et al[7] reported 3 of 7 cases of myxomas of the head and neck that had persistent diseases after incomplete resection. They also reviewed 169 cases from the English language literature and found that recurrence rates were 28 per cent for conservative surgery and only 6 per cent for local or wide excision with clear margins. Radiotherapy has been shown to have short-term benefit but no long-term cures by some authors[8,9], while others have reported that myxomas are not radiosensitive[2].

This reported case showed an extensive involvement of myxomas to the surrounding paranasal sinuses, orbits, and brain as reported earlier by some authors[6]. The treatment given at the first admission seemed to be inadequate resection. Recurrence occurred and readmission for the extensive neurosurgical and transnasal resection of the tumor was done. Although no residual tumor was detected at the early post-operative period, close follow-up was necessary because of their high recurrent rate.

SUMMARY

A case of myxoma of the nasal cavity and paranasal sinuses was presented. The extensive invasion of the tumor caused blindness and multiple operations. Complete resection is the treatment of choice while radiotherapy is not recommended.

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เนื้องอกมิย์กโซมาของจมูกและโพรงอากาศข้างจมูก : รายงานผู้ป่วย 1 ราย

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เนื้องอกมิย์กโซมาเป็นเนื้องอกที่เกิดจากเนื้อเยื่อเกี่ยวพัน (connective tissue) ประกอบด้วยเซลล์รูปดาวและกระสวย (stellate and spindle-shaped cells) ใน myxoid stroma เนื้องอกชนิดนี้พบบ่อยที่หัวใจ เนื้อเยื่ออ่อน (soft tissue) และกระดูก แต่พบได้น้อยมากที่จมูกและโพรงอากาศข้างจมูก ถึงแม้ว่าจะไม่ใช่มะเร็ง แต่มีการลุกลามไปสู่อวัยวะข้างเคียงได้สูง ทำให้การผ่าตัดออกให้หมดทำได้ยาก จึงเกิดการกลับเป็นซ้ำได้สูง รายงานฉบับนี้นำเสนอผู้ป่วย 1 ราย ที่เป็นเนื้องอกมิย์กโซมาของจมูกและโพรงอากาศข้างจมูก มีการลุกลามเข้าไปในสมอง และทำให้เกิดตาบอดทั้ง 2 ข้าง ผู้ป่วยได้รับการผ่าตัด 3 ครั้ง รวมถึงการผ่าตัดทางกะโหลกศีรษะ จึงสามารถควบคุมโรคไว้ได้

คำสำคัญ : มิย์กโซมา, จมูก, โพรงอากาศข้างจมูก

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