INTRODUCTION:

Meningiomas are one of the largest groups of brain tumors, which have two forms; intracranial and extracranial, extracranial location is very rare. Approximately 6-17% of all meningiomas can be found in extracranial regions [1].

Male patients are more common to have extracranial meningiomas [2].

CASE PRESENTATION

TREATMENT:

Total maxillectomy together with the excision of the tumor and the adjacent paranasal structures, following reconstruction of the orbit and maxilla with tissue patch was performed.

DISCUSSION

Meningiomas can exist as intracranial or extracranial brain tumors, which are benign, slow-growing tumors, extracranial location accounts for 2% of all these tumors [3] and found most often in male patients and in young individuals [4]. Due to their unusual symptoms and lack of prevalence, primary extracranial meningiomas are often misdiagnosed [5], fortunately, 80% of extracranial tumors are benign [6]. Cases of extracranial meningioma of sinonasal tract [7], retromolar area [8], eyebrows [9], pelvis [10], etc. had also been reported. Some of the published reports of extracranial meningiomas are listed in the table (Table 1). Histologically, primary extracranial meningiomas do not differ from intracranial, and most of these tumors are sporadic and the etiology remains unclear [11]. Primary extracranial meningiomas have been considered as arising independently from cranial nerve sheaths or from extracranial embryonic rests of arachnoid cells, and as extracranial metastases of a primary intracranial meningioma, but their origin has not been completely established [12].

The present case shows the clinical and imaging aspects of extracranial meningioma of the maxillary sinus in an old lady. Primary extracranial meningioma of the paranasal sinuses is rare [13]. In general, the most common signs and symptoms of paranasal sinus meningiomas may mimic cases of sinusitis with nasal obstruction, anosmia, facial pressure or pain, epistaxis and rhinorrhea [14,15].

Clinical examination should be comprehensive because more than 10% of cases may remain asymptomatic even in advanced stages [15].

The differential diagnosis should include a variety of benign and malignant neoplasms such as melanoma, olfactory neuroblastoma, carcinoma, hemangioma, sarcoma and aggressive psammomatoid ossifying fibroma [10, 14].

Histology is therefore essential and the general histologic features and immunohistochemically findings can usually differentiate between these tumors, as extracranial meningioma present with solid nests of meningothelial cells arranged in sheets or whorls with a fibroadipose background [5,13].

External beam radiation therapy has been shown to be effective and therefore reserved as a palliative approach [16].

References:

1. El-Daly A, Pitman KT, Ferguson BJ, Snyderman CH. Primary extracranial meningioma of the maxillary antrum. Skull Base Surg 1997; 7: 211-215 [PMID: 17171033 DOI: 10.1055/s-2008-1058598]

2. Claus EB, Bondy ML, Schildkraut JM, Wiemels JL, Wrensch M, Black PM. Epidemiology of intracranial meningioma. Neurosurgery 2005; 57: 1088-95; discussion 1088-95 [PMID: 16331155 DOI: 10.1227/01.neu.0000188281.]

3. Friedman CD, Costantino PD, Teitelbaum B, Berktold RE, Sisson GA Sr. Primary extracranial meningiomas of the head and neck. Laryngoscope 1990; 100: 41-48 [PMID: 2104554 DOI: 10.1288/00005537-199001000-00010]

4. Ho KL. Primary meningioma of the nasal cavity and paranasal sinuses. Cancer 1980; 46: 1442-1447 [PMID: 7417944 DOI: 10.1002/1097-0142(19800915)46:6<1442:]

5. Nager GT, Heroy J, Hoeplinger M. Meningiomas invading the temporal bone with extension to the neck. Am J Otolaryngol 1983; 4: 297-324 [PMID: 6416092 DOI: 10.1016/s0196-0709(83)80018-0]

6. Suárez-Fente V, Llorente-Pendás JL, Gómez-Martínez J, García-González LA, López-Alvarez F, Suárez-Nieto C. [Primary tumours of the parapharyngeal space. Our experience in 51 patients]. Acta Otorrinolaringol Esp 2009; 60: 19-24 [PMID: 19268125]

7. Maharjan L, Neupane Y, Pradhan B. Primary Atypical Meningioma of the Nasal Cavity: A Case Report and Review of the Literature. Case Rep Otolaryngol 2018; 2018: 7541892 [PMID: 29682381 DOI: 10.1155/2018/7541892]

8. Rege ICC, Garcia RR, Mendonça EF. Primary Extracranial Meningioma: A Rare Location. Head Neck Pathol 2017; 11: 561-566 [PMID: 28401439 DOI: 10.1007/s12105-017-0813-2]

9. Lee DH, Sim HS, Hwang JH, Kim KS, Lee SY. Extracranial Meningioma Presenting as an Eyebrow Mass. J Craniofac Surg 2017; 28: e305-e307 [PMID: 28212124 DOI: 10.1097/SCS.0000000000003555]

10. Nur S, Chuang L, Ramaswamy G. Primary extracranial meningioma of the pelvis: a light microscopic, immunohistochemical, and ultrastructural study. Gynecol Oncol 2006; 103: 745-748 [PMID: 16904168 DOI: 10.1016/j.ygyno.2006.05.050]

11. Lingen MW, Rao SM, Hutten MC, Pelzer HJ. Primary ectopic meningioma of the maxillary sinus: case report and review of the literature. Head Neck 1995; 17: 258-262 [PMID: 7782212 DOI: 10.1002/hed.2880170315.]

12. Takeshima Y, Kaneko M, Furonaka O, Jeet AV, Inai K. Meningioma in mature cystic teratoma of the ovary. Pathol Int 2004; 54: 543-548 [PMID: 15189511 DOI: 10.1111/j.1440-1827.2004.]

13. Arias Marzán F, de Lucas Carmona G, Alvarez Flórez M, Febles García P. [Extracranial meningiomas of the paranasal sinuses]. Acta Otorrinolaringol Esp 2010; 61: 238-240 [PMID: 20452881 DOI: 10.1016/j.otorri.2009.05.003]

14. Nozaki S, Yamazaki M, Koyama T, et al.: Primary extracranial meningioma of the maxillary sinus presenting as buccal swelling. Asian Journal of Oral & Maxillofacial Surgery 23: 134-137, 2011. [DOI: 10.1016/j.ajoms.2011.02.006]

15. Swain RE Jr, Kingdom TT, DelGaudio JM, Muller S, Grist WJ. Meningiomas

of the paranasal sinuses. Am J Rhinol 2001; 15: 27-30 [PMID: 11258651 DOI: 10.2500/105065801781329419]

16. Albsoul N, Rawashdeh B, Albsoul A, Abdullah M, Golestani S, Rawshdeh A, Mohammad M, Alzoubi M. A rare case of extracranial meningioma in parapharyngeal space presented as a neck mass. Int J Surg Case Rep 2015; 11: 40-43 [PMID: 25912007 DOI: 10.1016/j.ijscr.2015.04.012]

FIGURE LEGENDS

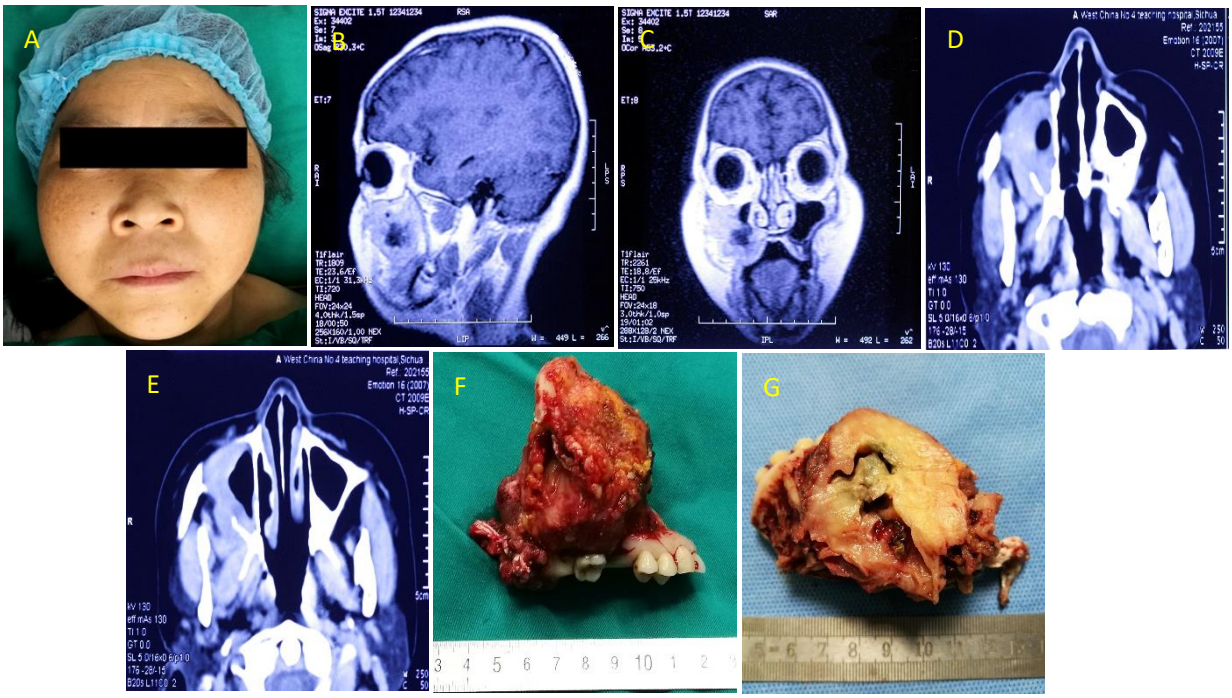

Fig 1 Characterization of imaging studies and gross finding.

A: Facial swelling measuring about 4 cm in diameter on right side;

B: MRI (sagittal section) demonstrating a soft tissue mass with a necrotic center compressing adjacent structures;

C: MRI (coronal section) demonstrating a soft tissue mass with a necrotic center compressing the right maxilla;

D, E: CT scan demonstrating a soft tissue mass with a necrotic center compressing adjacent structures along with the anterior wall of right maxilla;

F: The mass appeared to be lobulated and yellow-white measuring about 8 cm in diameter;

G: On hemisection, the mass showed well-circumscribed heterogenous lesion with a necrotic center.

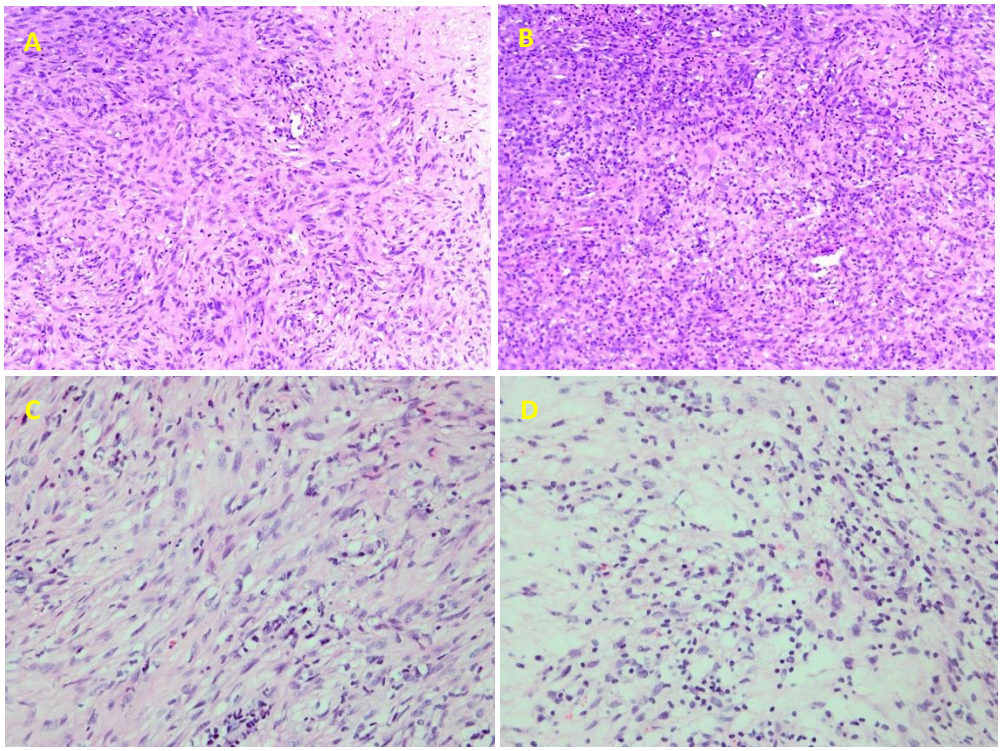

Fig 2 Histological features of extracranial meningioma.

A: The specimen showed epithelioid lobulated tissue, separated by abundant collagen fibers (H&E 100x).

B: The specimen showed epithelioid lobulated tissue, separated by abundant collagen fibers (H&E 40×).

C: The specimen showed abundant cytoplasm and indistinct cytoplasmic borders, arranged in whorled and lobulated patterns (H&E 100×).

D: The specimen showed abundant cytoplasm and indistinct cytoplasmic borders, arranged in whorled and lobulated patterns (H&E 40x).

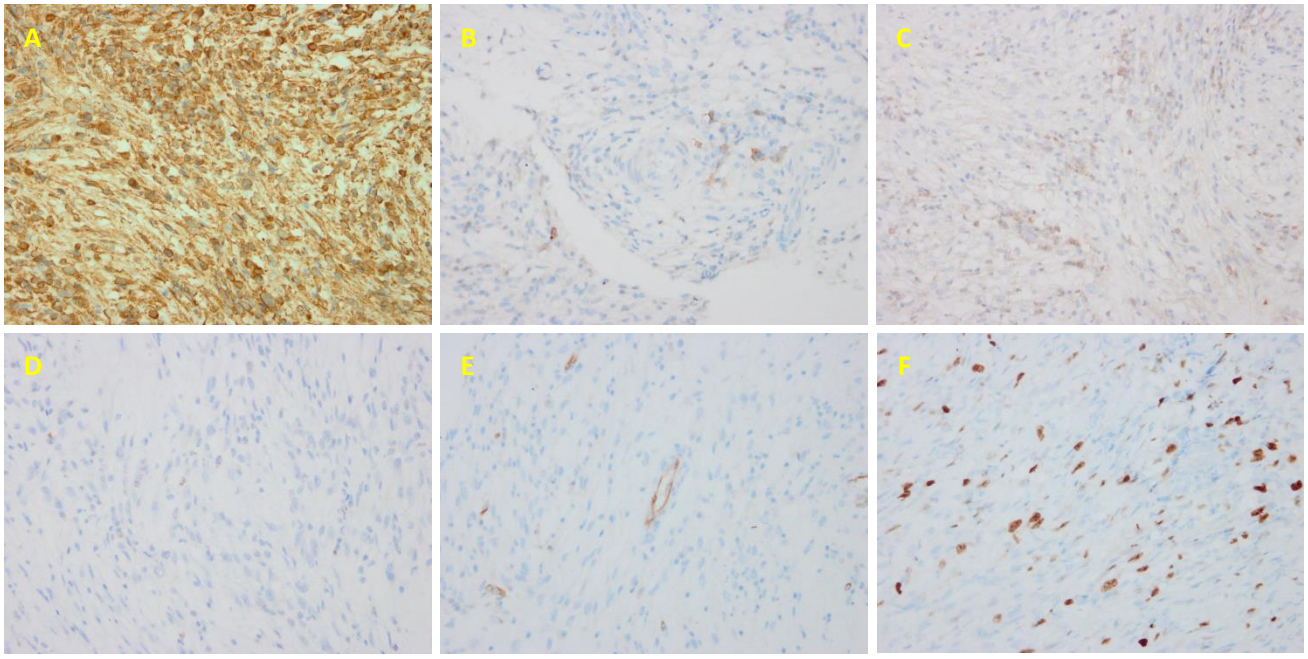

Fig 3 Immunohistochemical findings of the lesion.

The tumour cells were:

A: strongly positive for vimentin;

B: focally positive for EMA;

C: focally positive for CD99;

D: negative for STAT6;

E: negative for CD34;

F: focally positive for Ki-67.

Table 1: Published case reports of primary extracranial meningioma.

Case no.	First author	Year of publication	Site of primary extracranial meningioma	Diagnostic tests	Histology	Treatment performed
1.	Leison Maharjan	2018	Nasal cavity	CECT scan	WHO grade II atypical transitional meningioma	Endoscopic excision of the mass.
2.	Chae Min Kim	2018	Forehead	CT scan	Lobular architecture composed of tumor cells with eosinophilic cytoplasm and indistinct cell border	Excisional biopsy under local anaesthesia
3.	Ahmed El-Daly	1997	Maxillary antrum	CT scan	Interlacing bundles of bland-appearing spindle cells associated with calcific deposit	Medial maxillectomy with complete removal of the tumor
4.	Khang-Loon Ho	1980	Right nasal cavity	Sinus x-ray and CT scan	Clearly demarcated meningioma with fibrous capsule and well-preserved pseudostratified respiratory epithelium	Ablation of the right frontal sinus, external ethmoidectomy. and excision of the right middle turbinate
5.	Samina Nur	2006	Right pelvic cavity	Pelvic sonogram	Lobulated pattern composed of solid sheets of tumor cells separated by connective tissue septae	Exploratory laparotomy with optimal debulking of the pelvic tumor
6.	Nader Albsoul	2015	Right side neck mass	CT scan and MRI	Meningothelial cells with intranuclear inclusion and	Partial excision of the mass

					multiple psammoma bodies.	
7.	Yukio Takeshima	2004	Right ovary	Abdominal CT scan	Mature cerebral tissue was also noted. Melanocytes with black pigment were scattered in the peripheral region of the brain tissue	Right salpingo-oophorectomy
8.	Mark W. Lingen	1995	Right maxillary sinus	CT scan	Bundles of ovoid and spindle-shaped cells arranged in broad bands	Total maxillectomy
9.	Inara Carneiro Costa Rege	2017	Right retromolar area	CBCT scan	Spindle cell neoplasm, without evidence of atypia, whorls suggesting meningothelial origin	Partial resection of the mandible and reconstruction with autogenous iliac tricortical bone
10.	Do Hun Lee	2017	Left eyebrow	CT scan	Tumor cells arranged in sheets or whorls, with occasional psammoma bodies	Surgical excision
11.	Krishna Sigdel	(Present case)	Maxillary sinus	CT scan and MRI	Epithelioid lobulated tissue, separated by abundant collagen fibers	Total maxillectomy with excision of tumour