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A case of superficial angio-mucinous tumour occurring in the nasal septum and review of the literature

Yan Ma[1], Qingfeng Yong[1], Yan Jiang[2], Yuping Ma[2]

[1]Gansu University of Traditional Chinese Medicine, Lanzhou, 730000, China;

[2]Gansu Provincial Hospital of Traditional Chinese Medicine, Lanzhou, 730000, China

ABSTRACT

Superficial angiomyxoma is a rare, high-recurrence, slow-growing, painless benign tumour of the superficial skin that is an encapsulated, soft, lobulated cystic tissue with most superficial blood Myxomas are solitary. soft, lobulated cystic tissue with most superficial blood Myxomas are solitary. Its multiple sites are in the head and neck, trunk, and limbs. It is rare to occur in the nasal septum. Here is a report of a patient with superficial angiomyxoma occurring in the nasal septum. The clinical manifestations were The clinical manifestations were headache, dizziness, and nausea. Paranasal sinus CT showed abnormal density on the right side of the posterior part of the nasal septum. Under general anesthesia, endoscopic nasal tumor resection was performed on the right side of the nasal cavity. The postoperative pathological diagnosis was superficial angiomyxoma. After the postoperative follow-up visit, the wound healed well and is currently being followed up. After the postoperative follow-up visit, the wound healed well and is currently being followed up.

KEYWORDS

Superficial angiomyxoma; Nasal septum; Surgical operation

1 Case report

The patient, female, 48 years old, was admitted to the Department of Neurosurgery of Gansu Provincial Hospital of Traditional Chinese Medicine on 16 March 2023 with "insufficient blood supply to the vertebral basilar artery". She was admitted to the Neurosurgery Department of Gansu Provincial Hospital of Traditional Chinese Medicine on 16 March 2023 for "insufficient blood supply to the vertebral basilar artery". The patient had episodic dizziness and nausea for 3 years. Before admission, dizziness, chest tightness, shortness of breath, and nausea had worsened significantly, and were slightly relieved after rest. Specialist examination showed that the patient had clear consciousness, self-opening eyes, tangential answers, co-operative examination, GCS=E4V5M6=15 points. There was no gaze in both eyes, and light response and collecting reflexes were present. There was no obvious abnormality in general superficial and deep sensation. Muscle strength of all four limbs was grade 5, and muscle tone was normal. Bilateral babesiosis was negative. There was no neck resistance and meningeal irritation was not elicited. The patient was referred to our department after completing the relevant examinations, and the CT of paranasal sinuses showed abnormal density of the right side of the posterior nasal septum, which was combined with enhancement, and was considered to be a tumourous lesion in the nose, and neurosurgical diseases were ruled out. The patient was referred to our department because he had no history of cardiac mucinous tumour, pigmented skin lesions or endocrine abnormalities. Specialist examination

showed localised swelling of the skin on the lateral side of the right nasal septum, with a tough texture and no tenderness. Nasal endoscopy showed bilateral nasal mucosal oedema and an elevated mass at the right posterior end of the nasal septum, with a smooth surface and a size of 3* 2.7*1.2 cm. After communicating with the patient and his family members, it was decided to perform a general anaesthesia nasal endoscopic resection of the right nasal cavity, during the operation, it was seen that the mass was adhered to the turbinate and the middle turbinate with a lack of clarity of boundaries, and the mass had a wide tip, so part of the middle turbinate was resected, and the septal dissection was performed by removing the inferior turbinate bone. The swelling was fully exposed, the peritoneum was incised, and the swelling was cauliflower-like, brittle, with rich blood supply and unclear boundary with the surrounding tissues. The swelling was gradually peeled off, and it was seen that the swelling involved the posterior end of the nasal septum, the anterior and inferior wall of the pterygoid sinus, and the adjacent bone was thinned by the compression. Postoperative pathological findings were returned: superficial angiomyxoma (SA), immunohistochemical staining: CD34 (vascular +), CK-p (-), Desmin (-), S-100 single (-), SMA (+), ki-67 (+2%).

2 Talk over

Superficial angio-mucinous tumours were first described by Carney et al. in 1985.[1] et al. in 1985, and is thought to be associated with an autosomal dominant disorder known as the "Carney complex", which is caused by mutations in PRKAR1A.[2] The Carney complex is caused by mutations in PRKAR1A and disorders in the PKA signalling pathway of cyclic AMP-dependent protein kinase.[3] It is caused by mutations in PRKAR1A and disorders in the PKA signalling pathway of cyclic adenosine phosphate-dependent protein kinase and has a variety of clinical manifestations, including cardiac mucinous tumours, facial freckles, epithelioid blue nevi, endocrinopathies, and trachomatous melanotic nerve sheath tumours. [4] The presence of hyperpigmented skin and mucosal lesions is an important hallmark of this complex. When a patient presents with an abnormally active endocrine secretion and exhibits SA in multiple parts of the body, as a physician one should consider the possibility of Carney's complex, and a thorough history, physical examination and cardiac ultrasound are essential. In 1988 Allen[5] et al. first defined superficial angio-mucinous tumour through 30 of 28 patients. Superficial angiomyxoma is a rare, benign, non-invasive mucinous soft tissue tumour located superficially[6] Superficial angiomyxoma is a rare, benign, non-invasive, mucinous soft tissue tumour located superficially, usually on the trunk, head or neck, and occasionally on the lower limbs and genitals.[7] It is usually found on the trunk, head or neck, and occasionally on the lower limbs and genitals. It is common in middle-aged men. Clinical manifestations are skin papules, nodules or greyish-white polypoid lesions[8] that are not ulcerated and are sometimes papillomatous. Size is variable, but usually between 1-5 cm[7]. The tumour usually grows slowly and painlessly, is locally invasive and locally recurrent, with a reported recurrence rate of 30-40%, without any metastatic potential.[10] The pathogenesis of SA is unknown and diagnosis is often difficult due to the lack of distinctive features such as fibroepithelial polypoid masses. However, superficial angio-mucinous tumours are characterised by a prominent mucus-like stroma and abundant vascularity, and the lesion remains superficial without affecting deeper structures. Pathological examination is the gold standard for the diagnosis of angio-mucinous tumours, but the diagnosis is usually made only after postoperative pathological examination of the surgically excised mass. In this case, a nasal mass was considered preoperatively, and the diagnosis was only clarified postoperatively by sending a pathology biopsy. The patient's headache, dizziness, and nasal congestion were significantly reduced after surgery, and the symptoms were considered to be

caused by the previous mass compressing the tissue.

3 Diagnosis and Differential Diagnosis

The diagnosis of this disease mainly relies on histopathological examination. Microscopically, the main manifestation of this disease is a multilobular growth pattern, the interstitium contains more mucus and thin-walled blood vessels with a little inflammatory cell infiltration, the tumour cells are spindle and stellate shaped, no heterogeneity, and there is no nuclear schizophrenic image.[9] The tumour cells were spindle and stellate in shape with no heterogeneity. Immunohistochemical staining showed that the tumour cells expressed Vimentin and CD34, but not S-100 and Desmin.[12] Immunohistochemical staining The presence of interstitial inflammatory cells, especially neutrophils, is important for its diagnosis. Clinically, it can be distinguished from aggressive angio-mucinous tumour, cutaneous mucinosis, cutaneous mucinous cyst, mucinous fibrosarcoma, and dermal nerve sheath mucinous tumour.[13] The following are some examples of such tumours. Aggressive angio-mucinous tumour: a rare, locally aggressive but non-metastatic soft tissue tumour[14]. It is a rare locally invasive but non-metastatic soft tissue tumour that is usually large and deep and occurs in adult females. It appears in the genital and pelvic region and is prone to local recurrence. Microscopically, they appear as poorly defined, lightly stained spindle-shaped cells and deeply stained small nuclei with numerous thick-walled blood vessels and occasional foci of vitreous degeneration and haemorrhage, and should be differentiated from SA occurring in the genital tract.[15] . Although both superficial and invasive angio-mucinous tumours carry a risk of recurrence, especially if incomplete excision is performed, in the case of invasive angio-mucinous tumours, excision alone is not sufficient. Due to its unique local invasive potential and more distant metastases, it must be a wide local excision[16]. Mucinous tumours all consist of a mucinous stroma containing spindle filaments, stellate and plump oval cells, but the composition and number of blood vessels contained in the tumour vary. Cutaneous mucinosis and cutaneous mucinoma-like cysts contain fissures and pools of mucus, are virtually avascular, and are seen as small, ill-defined nodular lesions <2 cm in diameter[17]. Mucinous fibrosarcoma: microscopic manifestations include distinct mucinous areas, scanty cells in the form of spindle and stellate shapes, marked vascular proliferation, and inflammatory cell infiltration, which are similar to those of SA. However, mucinous fibrosarcoma is not common in superficial parts of the skin, with more obvious nuclear heterogeneity and increased chromatin, and the presence of curved blood vessels arranged by cells with deeply stained nuclei.[18]. Dermal nerve sheath mucinous sarcoma: It occurs in the dermis and subcutis, and is broadly similar to mucinous sarcoma. Microscopically, they appear as well-defined, lobulated components with fibrous tissue segmentation and mucus-like interstitium. The cells are benign, spindle-shaped arranged in fascicles and spirals with mild anisotropy and nuclear division. However, unlike SA, there is no inflammatory cell infiltration and abundant vascularity and S-100 protein-negative reaction within the tumour cells[19]. Other differential diagnosis should also be made with epidermoid cysts, lipomas, neurofibromas, abscesses, lymphangiomas, and dermatofibromas.

4 Treatment and prognosis

SA is a benign tumour and the treatment of choice is surgical wide excision of the[20]. Even with this surgical approach there is still a possibility of recurrence due to confusing clinical margins and incomplete surgical resection.[21] However, recurrences tend to be localised, non-destructive and without distant metastases, so the prognosis for SA is good, but due to the high rate of recurrence

regular review and follow-up is required.

About the Author

Yan Ma, female, 1998, postgraduate student of Gansu University of Traditional Chinese Medicine, Class 2021. Her research interest: Integrated Chinese and Western Medicine for Ear, Nose and Throat Diseases, E-mail: 2286230159@qq.com;

Qingfeng Yong, male, 1992, mast's degree, attending physician research direction: Integrated Chinese and Western Medicine for Ear, Nose and Throat Diseases, E-mail: 931381078@qq.com;

Yan Jiang, female, 1974, bachelor's degree, chief physician, postgraduate supervisor, research direction: Integrated Chinese and Western Medicine for Ear, Nose and Throat Diseases, E-mail: 1754943546@qq.com.

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