Aggressive sinonasal angiomyxoma management with endoscopic medial maxillectomy approach: a rare case report

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INTRODUCTION

Aggressive angiomyxoma is a benign tumour of mesenchymal origin with a very rare incidence. Aggressive angiomyxoma often occurs, especially in the pelvic, vulvovaginal and perineal areas in women of childbearing age, while in men, it occurs in the reproductive organs in old age, with a ratio between women and men around 6:1. Only a few cases have been reported involving the extragenital area.²⁻⁵ Aggressive angiomyxoma is often found in reproductive age, with a peak in the age group of 31 to 35 years.³ Sinonasal angiomyxoma can develop in bone or soft tissue. The cause of sinonasal angiomyxoma is generally not known with certainty. The suspected etiologic factors are congenital abnormalities and the presence of hormonal influences.⁴

Clinically, due to its rarity and non-specific symptoms, early diagnosis is difficult and usually found on histopathological examination.¹,⁶⁻⁸ Symptoms and signs on head and neck examination are nasal obstruction, malocclusion, tooth instability, pain, and diplopia. The differential diagnosis of aggressive sinonasal angiomyxoma includes angiofibroma, sinonasal polyp, inverted papilloma (IP) and a malignant tumour if bone destruction is found.⁴ The diagnosis can be established based on clinical, radiological, and histopathological findings of mesenchymal tumours and the presence of fibroblasts against a myxoid background.³

The treatment of choice for aggressive sinonasal angiomyxoma is the extirpation of the mass with a surgical procedure according to the site of the tumour. Surgical procedures can be performed endoscopically or externally.⁶ Therefore this study aims to report the management of aggressive sinonasal angiomyxoma with an endoscopic approach. This aggressive sinonasal angiomyxoma was the first rare case found in Dr. Soetomo Hospital, Surabaya, Indonesia.

CASE PRESENTATION

A 20-year-old female came to the Otorhinolaryngology Head and Neck Surgery Outpatient Unit, Dr. Soetomo Academic General Hospital, Surabaya, Indonesia, on March 3, 2020. The patient was referred from Dr. Wahidin Sudiro Husodo Hospital, Mojokerto, Indonesia, and initially diagnosed with right antrochoanal polyp (ACP). In the previous six months, the patient had complained of intermittent nasal blockage since the patient also complained of frequent colds accompanied by clear, watery, odourless mucus. The patient often experienced intermittent nosebleeds from the right nose, with varying amounts, but they could stop spontaneously even though they took a longer time.

Since the previous month, the right had been persistently blocked, and sometimes, the left nose also felt stuffy. It felt like there was meat stuck in the nose. Nosebleeds became more frequent. The patient complained of intermittent epistaxis from the right nose, which could stop by itself but in a longer time. The patient’s case was indicated with the inclusion criteria and stated that aggressive sinonasal angiomyxoma is rare. Extirpation surgery was performed with an endoscopic medial maxillectomy approach. Histopathology showed a benign mesenchymal cell tumour consistent with angiomyxoma.

Background: Aggressive angiomyxoma is a benign tumour of mesenchymal origin, and its incidence is very rare. The treatment of choice for aggressive sinonasal angiomyxoma is surgical extirpation based on the location of the tumour. The surgical procedure can be performed endoscopically or externally. This study aims to report the management of an aggressive sinonasal angiomyxoma rare case with an endoscopic approach.

Case presentation: A 20-year-old female came with the chief complaint of a blocked right nose for the past six months. Patients often had varying amounts of intermittent epistaxis from the right nose, which could stop by itself but in a longer time. Three studies met the inclusion criteria and stated that aggressive sinonasal angiomyxoma is rare. Extirpation surgery was performed with an endoscopic medial maxillectomy approach. Histopathology showed a benign mesenchymal cell tumour consistent with angiomyxoma.

Conclusion: The sinonasal tumour with an early preoperative diagnosis of an antrochoanal polyp and a postoperative diagnosis of an aggressive sinonasal angiomyxoma underwent extirpation surgery with an endoscopic medial maxillectomy approach results in no recurrences after two years.

Keywords: Aggressive sinonasal angiomyxoma; endoscopic medial maxillectomy, tumour.
headache, especially in the forehead, the right cheek felt a little thick, there was an olfactory disorder in the right nose, and tinnitus in the right ear. There were no complaints of hearing loss, blurred vision or double vision, loose teeth, lumps on the neck, lumps in the armpits and groin. The patient had a history of trauma to the nose or previous nose surgery, a history of taking birth control pills, allergy, diabetes, high blood pressure, or another severe chronic disease.

A physical examination showed that the patient was in comos mentis condition and the vital signs were within normal limits. A deformity of the nasal dorsum was found, suggesting a mass pressing from the right nasal cavity (Figure 1a). The grey mass seemed to fill the right nasal cavity. The surface was flat, soft, slippery, seemed easy to bleed, there was no tenderness and minimal mucopurulent secretions. The left nasal cavity was within normal limits. There was a tender mass on the right side. The right side of the soft palate was bulging, not hyperaemic (Figure 1b). Examination of the ears and neck revealed abnormality. The outcome of the patient showed no recurrence after two years in which no cyst formation was found in nasal endoscopy post-operatively.

**DISCUSSION**

Aggressive angiomyxoma is a tumour of mesenchymal origin. Mesenchymal cells are star-shaped cells, smaller than fibroblasts, usually located along the walls of capillaries and are known as perivascular cells, adventitia cells or pericytic cells. Research by Cao et al., 2022 showed that aggressive angiomyxoma was more common in women, with a ratio between women and men of about 6:1. This tumour is often found in reproductive age with a peak at the age of 31 to 35 years. The sex of the patient in this case was in accordance with that in the literature, but at a younger age. This tumour is often found in the pelvic, vulvo vaginal, and perineal regions.

In this patient, a grey mass was found to fill the right nasal cavity, with a flat, soft, smooth surface, with the impression of being easy to bleed. This profile agreed with that in the literature, but the site of the mass was inconsistent. The etiology of sinonasal aggressive angiomyxoma is still unclear. The pathogenesis of hemangiomas is a neoplastic process. Study by Banasser et al., 2020 supporting the statement that the predisposing factors in this tumour were still unclear. It is suspected that the influence of the hormone estrogen is a factor that causes tumours. The aetiology of the disease in this patient was also unclear, but the influence of the hormone estrogen could not be ruled out. Transnasal Esophagoscopy is a small caliber flexible esophagoscopy technique performed transnasally to evaluate the esophageal lumen.

Early diagnosis of aggressive angiomyxoma sinonasal is difficult because it is a rare case, and the symptoms are not specifics, so its diagnosis is established histopathologically. Research by Lee at al., 2021 found that the symptoms of aggressive angiomyxoma sinonasal depend on the extent of the tumour. If the tumour involves the sinonasal area, the emerging clinical symptom is unilateral nasal obstruction which may be
Figure 3. Right nasal cavity. (a) The mass looked yellowish white smooth and bled easily, (b) The mass was extracted with Blakesley’s forceps, (c) Bleeding is controlled with gauze, (d) Incision of the lateral mucosa of the nasal cavity, (e) Chiseling the lateral wall of the nasal cavity, (f) Bleeding of the branch of a sphenopalatine artery (yellow arrow), (g) Removal of the tumour mass in the nasopharynx, (h) Removal of the tumour mass in the maxillary antrum, the tumour mass appears smoother, (i) The wound area was closed with Surgicel®.

Figure 4. Postoperative aggressive angiomyxoma mass.

accompanied by rhinorrhea or epistaxis. Secretions discharging from the nose are often mixed with blood. Large tumour size can cause nasal deformity due to the tumor pressing against bone. Davidson et al., 2021 reported an aggressive sinonasal angiomyxoma with similar symptoms, including nasal obstruction, anosmia, rhinorrhea and epistaxis. The complaints experienced by the patient, in this case, were in accordance with those in the literature on benign masses in sinonasal. The duration of surgery outcome showed significantly different results compared to previous outcomes.

Differential diagnosis in large lesions can lead to misdiagnosis of myxofibrosarcoma, rhabdomyosarcoma, melanoma and nasopharyngeal angiofibroma. The preoperative histopathology of this patient was ACP, and the postoperative histopathology was angiomyxoma. A definite diagnosis of aggressive angiomyxoma can be established through histopathological examination. The histopathological profile may consist of stellate and spindle cells embedded in a loose matrix of corrugated collagen and containing thin-walled capillaries and thick-walled vascular channels. The histopathology of the tumour in this patient was in accordance with that in the literature. Decompression surgery (left hemilaminectomy) and tumor extirpation were thereafter carried out on the patient after the routine laboratory test was performed.

The recommended supportive examinations are CT scans and magnetic resonance imaging (MRI). A CT scan shows an image of opacity in the nasal cavity accompanied by the narrowing of one or more paranasal sinuses and may be accompanied by bone destruction. The appearance and expansion of the mass were in accordance with those in the literature and the patient was not subjected to MRI because two biopsies had been performed. Management of sinonasal angiomyxoma was still controversial. The most recommended management is operative therapy. Cao et al., 2022 wrote that in principle, hemangiomas should be extirpated at all ages. If sinonasal angiomyxoma is associated with pregnancy, regression usually occurs after delivery.

Recurrence can occur if the tumour mass is not obliterated. The surgical approach used must be adapted to the location and size of the tumour. Surgical techniques can be performed endoscopically or externally, which is more radical. In this case, extirpation was performed with an endoscopic approach because the size of the tumour could be reached by endoscopy and the patient’s age was young in the hope of preserving the aesthetics of the patient. The difference in the surgical approach planned before and during the operation was due to the initial diagnosis of ACP. The complex endoscopic
endonasal surgery group required a longer length of care and more post-operative outpatient visit than the simple procedure group.\(^{15}\)

A two-year postoperative evaluation with nasal endoscopy revealed no recurrence of the tumour mass. Several studies and case reports have shown that recurrence is common when residues remain on the lesion’s surface. Buurman et al., 2020 suggested that recurrence was common in about 30-40% of cases and occurred from several months to several years after surgery.\(^{13}\) Research conducted by Singh et al., 2020 reported recurrence up to two years post-operatively.\(^{16}\) Aminimoghaddam et al., 2023 reported that recurrence occurred at four months and sixty months postoperatively.\(^{2}\) The most important thing to pay attention to is strict follow-up for possible recurrence of the tumor.\(^{17}\) Presently, there are no studies on the recurrence of aggressive angiomyxoma sinonasal.

**CONCLUSION**

The endoscopic medial maxillectomy technique was used to remove the sinonasal tumor which had been diagnosed as an aggressive sinonasal angiomyxoma post-operatively after an early preoperative diagnosis of an antrochoanal polyp with no recurrence after two years.

**CONFLICT OF INTEREST**

The author reported no conflicts of interest in this study.

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**ETHICAL CLEARANCE**

The authors have secured informed consent from the patient regarding this study.

**AUTHOR CONTRIBUTION**

All of the authors equally contributed to the study.

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Figure 5. Left nasal cavity. (a) Removal of crust and a blood clot in the left nasal cavity (yellow arrow), (b) Minimal oedema mucosa and crusting and cyst formation (blue arrow), (c) Cyst formation deflated with Blakesley forceps, (d) Visible fibrosis and granulation tissue, minimal crusting, and minimal cyst formation in the right maxillary antrum.