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Juvenile Nasopharyngeal Angiofibroma: Management Challenges in a Tertiary Health Institution in Sokoto, Northwestern Nigeria

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Authors' contributions

This work was carried out in collaboration among all authors. Author SBA designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors MA, DA, KRI, KA and UM managed the analyses of the study. Authors SUY and JCO managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Aims: To highlight the diagnostic and therapeutic challenges in the management of the patient.
Background: Juvenile nasopharyngeal angiofibroma is a rare, benign, and vascular tumour that occurs almost exclusively in male teenagers. The tumour usually originate in the posterior nasal cavity around the sphenopalatine foramen and nasopharynx. It is characterized by spontaneous, recurrent and life-threatening epistaxis, nasal and nasopharyngeal mass. Also, it is non-infiltrating but could spread into soft tissues, foramina, orbit, intracranial, and regress at secondary sexual maturity. We report a case of a 13-year-old boy with Juvenile nasopharyngeal angiofibroma complicated by upper airway obstruction and highlighted the diagnostic and therapeutic challenges in his management.

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Case Report: A 13-year-old Fulani teenager, presented with a one-year history of progressive bilateral nasal blockage, three months history of growth in the mouth, dysphagia, mouth breathing and epistaxis. Physical examination revealed a young boy in apparent respiratory distress, muffled voice, mouth breathing with rhinolalia clausa. Pinkish mass filled the right nasal cavity and nasopharynx. Computerized tomographic scan imaging showed a huge enhancing hyperdense mass occupying the nasopharyngeal, right nasal cavity and oropharyngeal airway. Had excision biopsy via combined lateral rhinotomy and transoral approaches under general anaesthesia. Histopathological studies confirmed the diagnosis of juvenile nasopharyngeal angiofibroma.

Conclusion: The patient had good result with invasive surgical intervention, despite, non-accessibility of CT angiography and embolization.

Keywords: Juvenile nasopharyngeal angiofibroma; challenges; Sokoto.

1. INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is a rare, benign, and vascular tumour that occurs almost exclusively in male teenagers [1,2]. JNA usually originate in the posterior nasal cavity around the sphenopalatine foramen, medial pterygoid plates, lateral wall of the nasopharynx, basilar part of the occipital bone, basisphenoid and the cervical vertebra [3,4]. Generally, JNA accounted for 0.5% of all head and neck tumours, and the global incidence is estimated at 1: 150,000 [1]. More than 400 cases were managed between 1990 and 2013 in India [5]. The available data of JNA in Africa is mainly from an isolated case report or case series. An earlier report [6] from Kenya reported five cases in 1964. In 1978, Martinson et al. [7] reported three cases involving one female and two male patients from Ibadan, Nigeria, and their age ranged from 13-16 years. Nine cases were reported in 1980 from Cape Town, South Africa [8]. One of the nine patients aged 42 years, and the remaining eight patients age range was 8-12 years, and they were all males. A case series from Cairo, Egypt [2] in 1981 reported 15 cases, and all were males with an age range of 12-22 years. Also, there was another case report [9] from Nigeria in 2013.

The aetiopathogenesis of JNA is many and controversial. The assumption includes arising from the periosteum of the skull base, hormonal factor, remnant of Cranio-pharyngeal duct, and misplaced sequestrated erectile tissue during gestation [1,2,5]. Clinically, JNA is characterized by spontaneous, recurrent and life-threatening epistaxis, nasal and nasopharyngeal mass which may push the soft palate downward into the oropharynx. Also, it is non-infiltrating but could spread into soft tissues, foramina, orbit, intracranial, and regress at secondary sexual maturity [1,10]. We report a case of a 13-year-old boy with JNA complicated by upper airway obstruction and highlighted the diagnostic and therapeutic challenges in his management.

2. CASE REPORT

13-year-old Fulani teenager referred from a tertiary hospital in Bauchi, northeastern Nigeria. He presented with a one-year history of progressive bilateral nasal blockage, three months history of growth in the mouth, dysphagia, mouth breathing, anterior and posterior epistaxis. Nasal blockage commenced at the right side and later involved the left nasal cavity. Epistaxis was spontaneous and recurrent over two months. Estimated blood loss was 500 mls. No bleeding from other orifices. The patient could not take both liquid and solid diet, and there was associated weight loss. Had difficulty in sleeping at night because of noisy breathing, disruptive snoring, and apneic episodes during sleep.

Physical examination revealed a young boy in apparent respiratory distress, muffled voice, mouth breathing with rhinolalia clausa. Pinkish mass filled the right nasal cavity and pushed the nasal septum to the lateral wall of the left nasal cavity. The soft palatal bulge and the pinkish mass descended behind the soft palate and almost abutting on the base of the tongue (Fig. 1).

Vital signs (respiratory rate 28/minute, temperature 37.2° centigrade, pulse rate 98/minute, and blood pressure 100/70 mm Hg). Chest, cardiovascular system, gastrointestinal system, and central nervous system were essentially normal.

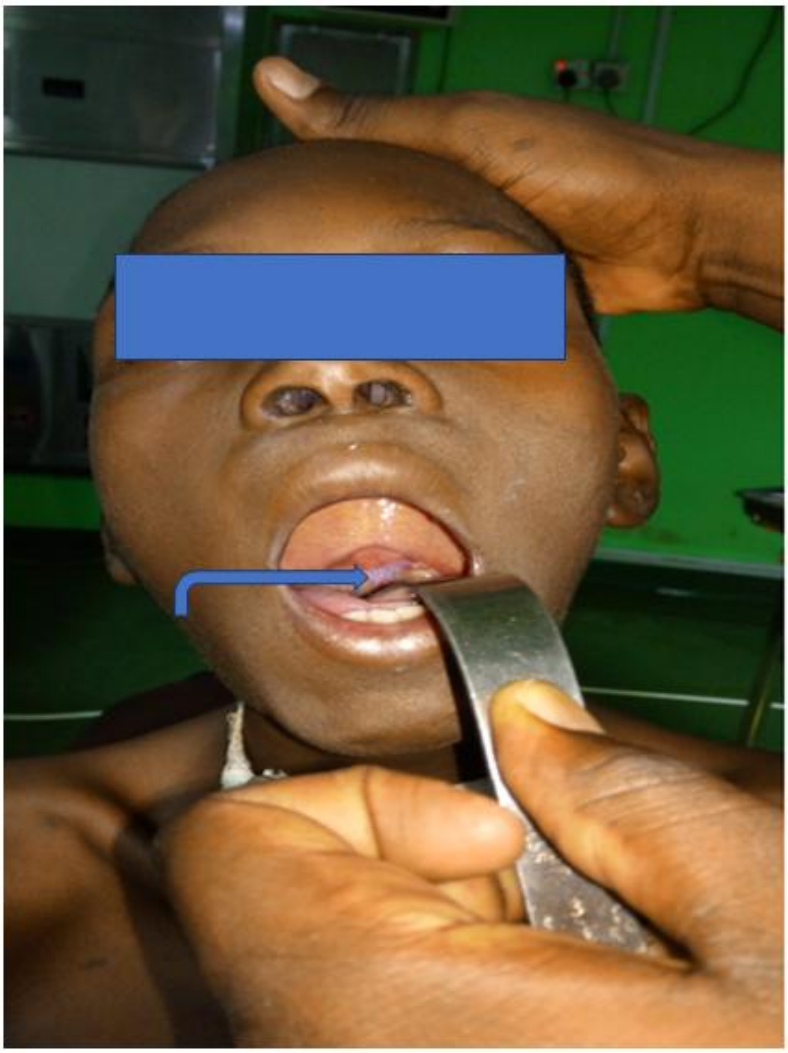

Fig. 1. Palatal bulge and pinkish mass descending into the oropharynx (blue arrow)

Computed tomographic (CT) scan imaging, scanogram shows haziness at the region of the nasopharyngeal and oropharyngeal air column as well as the right nasal cavity. Serial axial slices and bone window in pre and post-contrast series done at 2.5mm intervals showed a huge enhancing hyperdense mass (HU; pre=41, post=86) occupying the nasopharyngeal and oropharyngeal airway and extend into the right nasal cavity. There is associated compression and displacement of the nasal septum to the contralateral side with bony erosion. Mucosal thickening bilaterally in the maxillary and ethmoidal sinuses (Figs. 2 and 3). Magnetic resonance imaging (MRI) and embolization were not done due to logistic problems.

Full blood count showed anaemia (packed cell volume=26%); other parameters were within a reasonable limit. Serum electrolytes, urea, and creatinine were within the standard limit, and chest X-ray was essentially normal.

He had an emergency tracheostomy, which was complicated by right-sided pneumothorax and right lung collapse. The pneumothorax resolved with closed thoracostomy tube drainage. Anaemia was corrected by transfusion of one pint of blood. He had definitive surgery via the right lateral rhinotomy and transoral approaches.

Haemostasis controlled by pressure packing with gauze and application of surgicel to the site of origin. Right external ethmoidectomy and inferior meatal intranasal antrostomy done, followed by posterior and anterior nasal packing with gauze. Moore incision closed in two layers. Subcutaneous layer with vicryl 3/0 and skin with nylon 4/0.

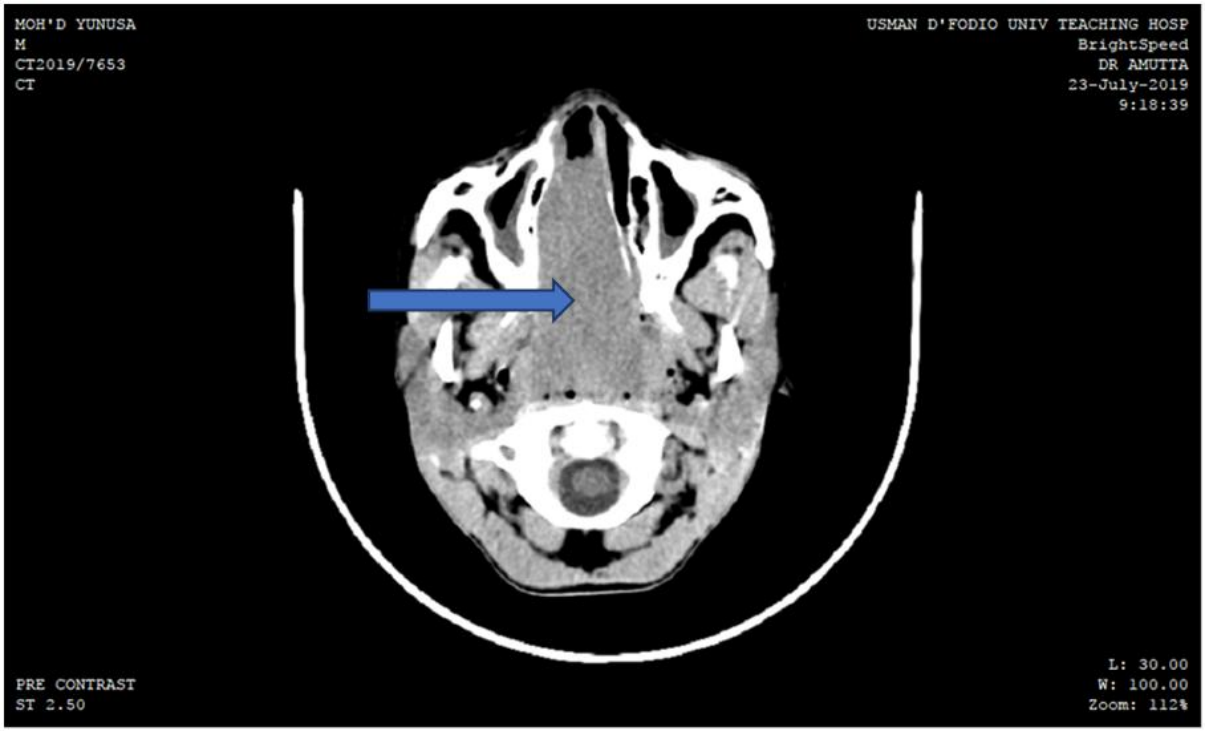

Fig. 2. Pre-contrast CT scan of the paranasal sinuses showing a huge isodense mass (blue arrow) in the nasopharynx and right nasal cavity

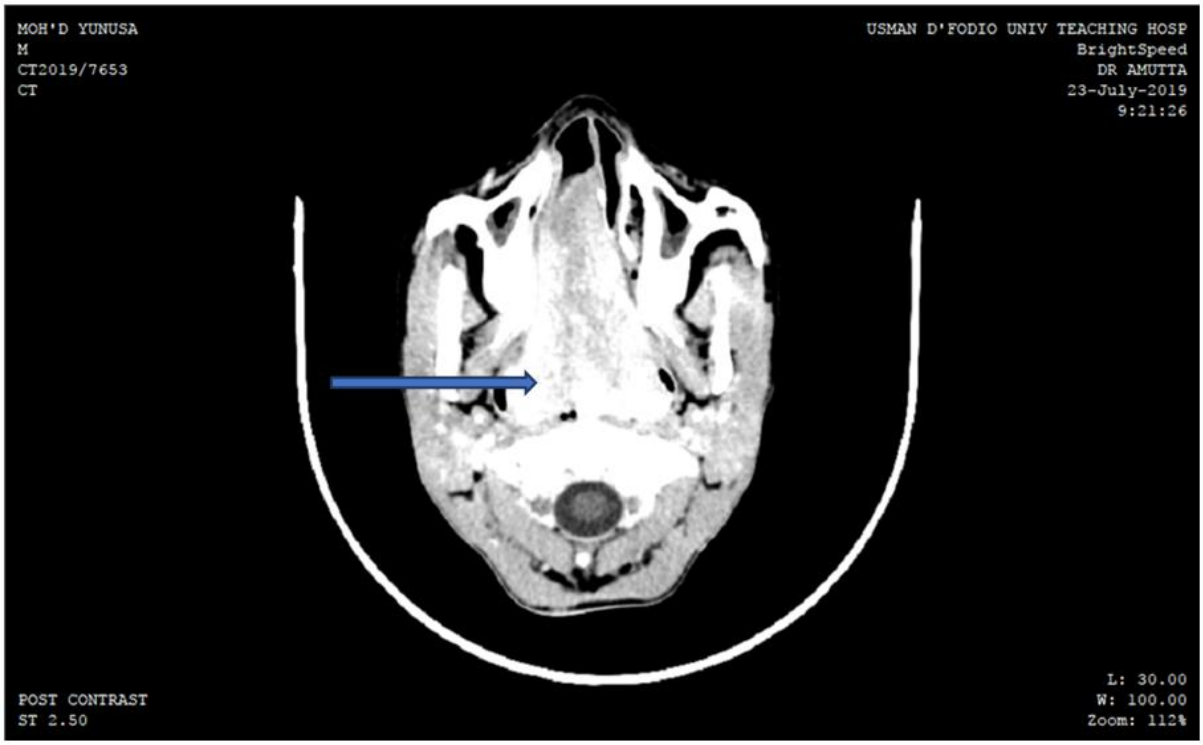

Fig. 3. Post-contrast CT scan of the paranasal sinuses showed a contrast-enhanced mass (blue arrow) in the nasopharynx and right nasal cavity

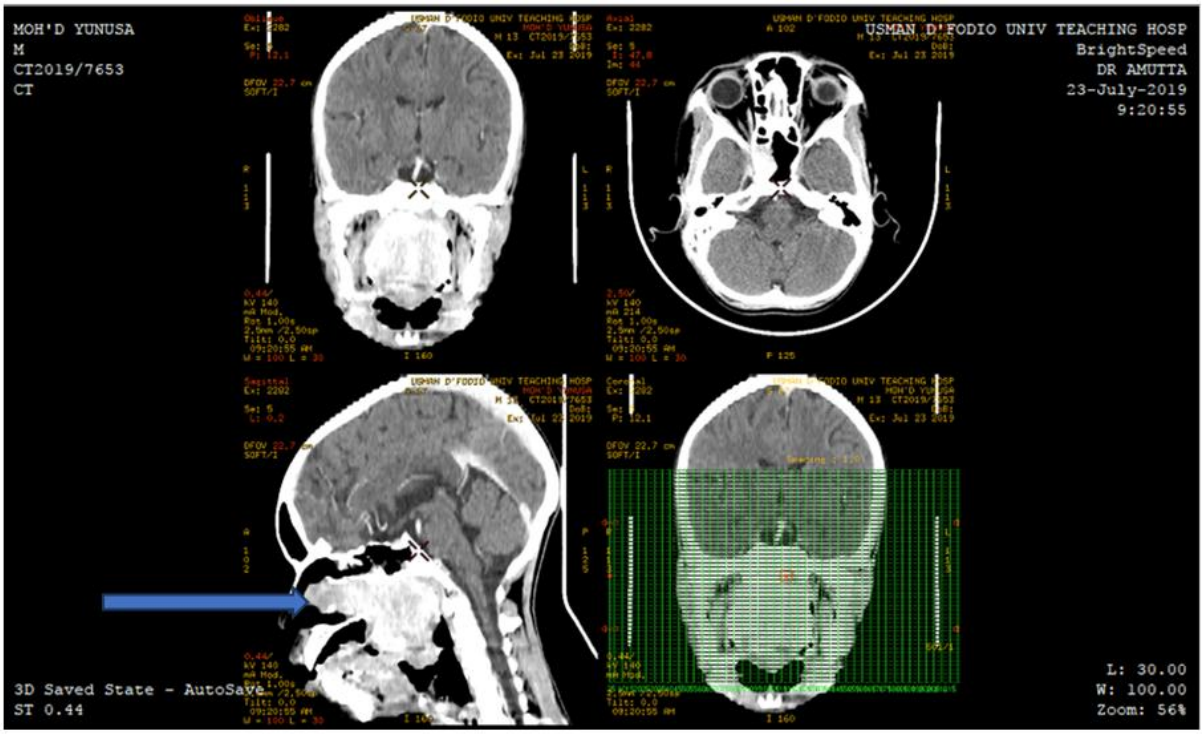

Fig. 4. Axial, coronal, and sagittal reconstruction of the CT scan paranasal sinuses showing enhancing mass (blue arrow) in the nasopharynx and nasal cavity

Operative findings were a substantial haemorrhagic mass arising from the right postero-lateral wall of the nasopharynx and extending into the right nasal cavity. It was removed by a combination of blunt and sharp dissection and finally delivered through the mouth. The excised specimen weighed 76g and measures 8 x 7 x 5 cm (Fig. 5).

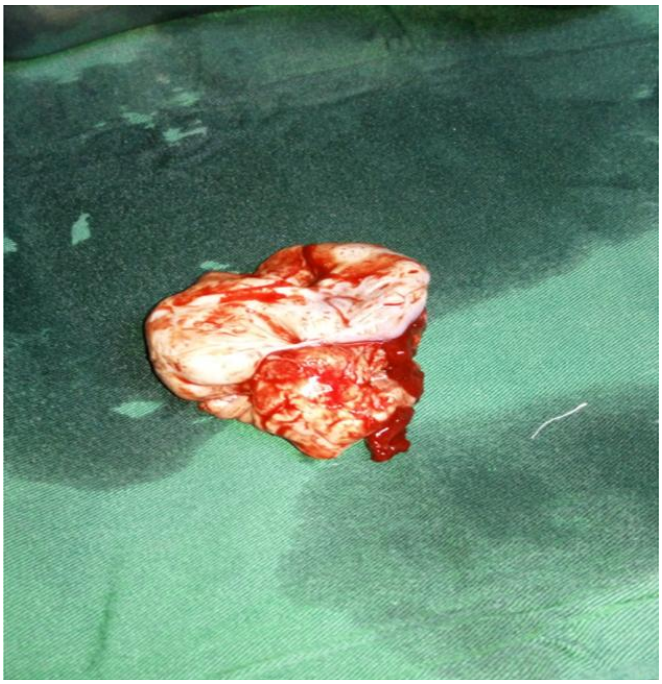

Fig. 5. Surgical specimen from the nasopharynx and right nasal cavity

Histopathological studies showed ulcerated polypoid tumour of a complex mixture of stellate fibroblast with bland nuclei and varying size of double-layered interconnecting to jagged vascular channels. Varying degree of collagenization in the interstitium, which is also sprinkled with mass cells. Conclusion: Nasopharyngeal angiofibroma (Fig. 6). However, immunohistochemical analysis was not performed to characterize the juvenile nasopharyngeal angiofibroma in this case study.

Postoperative treatment consisted of intravenous fluid with 5% dextrose saline 500 ml 6-hourly for 24 hours. Intravenous ceftriaxone (Avicef) 1g 12-hourly for 72 hours, intravenous metronidazole 500mg 8-hourly for 72 hours and intravenous paracetamol 450mg 8-hourly for 24 hours. Oral feeding was commenced on the first day postoperative period. Intravenous antibiotics were converted to tablet Augmentin 625 mmg 12-hourly and metronidazole 400mg 8-hourly on the 4th postoperative day. Tracheostomy was decannulated on the 4th postoperative day and sutures removed on the 7th day after surgery. He was discharged home on the 9th postoperative day in excellent clinical status

3. DISCUSSION

The treatment of Juvenile nasopharyngeal angiofibroma has undergone constant evolution. Early surgical intervention at the time of Celsus (2nd-century Greek philosopher) was by using a snare, digital manipulation and tearing the tumour with forceps, followed by triple therapy consisting of sex hormone therapy, radiation therapy and surgery from 1927 to 1947 [10]. Effective therapeutic options currently applied include surgery, radiotherapy, and in rare cases, chemotherapy [4].

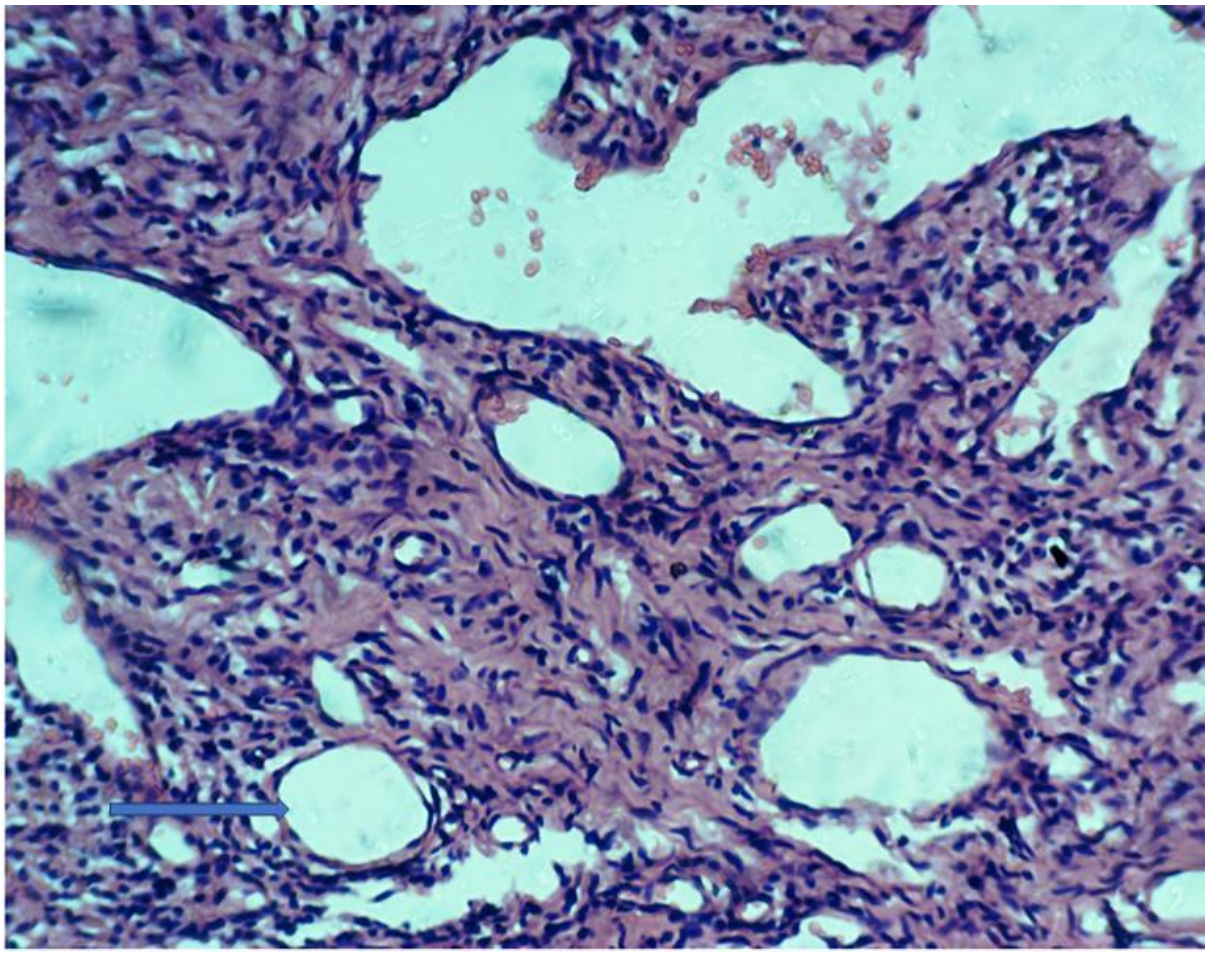

Fig. 6. Photomicrograph showing thin-walled and large vascular channels (blue arrow) in the fibrous stroma (H and E x200)

Surgical resection of JNA is done by many techniques ranging from open surgeries via facial degloving, midline skin incision like lateral rhinotomy and lately endonasal endoscopic resection in selected cases [4,11]. A pre-requisite for limiting life-threatening haemorrhage, during surgery, is identifying JNA feeding vessels. Maxillary and sphenopalatine arteries are the most common feeding vessels. Additionally, the ascending pharyngeal artery and less commonly from branches of the internal carotid and vertebral arteries. Therefore, a CT scan imaging and CT angiography with selective embolization of JNA feeding arteries is the established practice among most surgeons [4,11,12]. The absence of CT angiography and embolization was a severe diagnostic challenge in the management of the patient in this case report. CT angiography and embolization are not available in the centre where this patient was managed and could not afford referral to another hospital because of financial constraint. Moreover, he was referred to our health institution from another tertiary hospital, where these facilities are also unavailable. In our opinion, there is a strong need to provide these services by the government and financially able individuals to aid the management of patients with similar conditions.

The main therapeutic challenge in the management of the patient in this case report was the choice of surgical intervention. The endonasal endoscopic approach is applicable with the superior cosmetic result since the JNA was confined to the nasal cavity, nasopharynx, and oropharynx [13]. Endoscopic excision has an advantage over the lateral rhinotomy by avoiding middle facial incision, the elevation of soft tissue, periosteum, and osteotomies that have the potential to induce abnormal facial bone growth in teenagers [14-16]. The choice of the combined lateral rhinotomy and transoral approach was due to lack of facility and technical experience for endoscopic resection of JNA. Nevertheless, similar approaches have been utilized with a good result by some surgeons [4,11].

The emergency tracheostomy performed for the index patient was due to severe upper airway obstruction. Comparatively, earlier reports [16] recommended it. Postoperative MRI is recommended to be carried out within 72-hours of JNA excision and every 6-8 months for three years [1]. The MRI to be done at follow-up is difficult in most developing countries, including Nigeria, because of no- availability of the MRI machine, and the high cost where it is available.

Radiotherapy is reserved for JNA with intracranial extension, patient's refusal to undergo surgery or unfit due to co-morbidities and residual disease after surgery [14,16,17]. Radiotherapy was not considered for the index case because examination of the posterior nasal cavity and nasopharynx showed complete tumour excision.

The patient in this report did well with the surgery and supportive medical treatment. The presenting symptoms of nasal blockage, rhinolalia clausa, epistaxis, mouth breathing, and dysphagia resolved entirely in the postoperative period. He was discharged home in excellent condition and will be monitor on follow-up for JNA recurrence.

4. CONCLUSION

The patient had good result with invasive surgical intervention, despite, non-accessibility of CT angiography and embolization. Provision of CT angiography, embolization and facilities for endonasal endoscopic surgery are required for good management of JNA.

CONSENT

Written permission was obtained from the father of the patient and kept by the authors.

ETHICAL APPROVAL

As per international standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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