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Diagnosis and management of juvenile nasopharyngeal angiofibroma

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Abstract
Juvenile nasopharyngeal angiofibroma (JNA) is a benign vascular tumour in the posterior part of the nasopharynx and sphenopalatine foramen. It is rare; its incidence only accounts for 0.05% to 0.5% of all head and neck tumours. Even though it is benign, JNA can be life-threatening because of its very aggressive nature, in which 10-37% of cases invade cranial and orbital.
One case of JNA was reported in a 13-year-old pediatric patient at Prof. Dr. I. G. N. G. Ngoerah Hospital Denpasar, Bali, Indonesia. The patient underwent a lateral rhinotomy with a left and trans-palatal midfacial degloving approach, where intraoperatively, a tumour mass that extended to the left nasal cavity, maxillary sinus, ethmoid and left sphenoid was found in the nasopharynx. In this case, we found no postoperative complications. This case is reported to provide information on the management of JNA.

Keywords: juvenile nasopharyngeal angiofibroma, pediatric, lateral rhinotomy, trans-palatal

Introduction

Juvenile nasopharyngeal angiofibroma (JNA) is a benign vascular tumour in the posterior part of the nasopharynx and sphenopalatine foramen. It is very scarce that the incidence of this tumour only accounts for 0.05% to 0.5% of all head and neck tumours. The prevalence of JNA in the United States is estimated to be only 0.4 per million population. Although it can occur at any age, the highest incidence is found in male patients aged 7 to 29 years [1, 2]. Because this disease is quite rare, there is no data regarding the epidemiology of JNA in Indonesia.

Even though it is benign, JNA can be life-threatening because of its very aggressive nature, in which 10-37% of cases involve cranial and orbital. This invasion can cause various morbidities, including nasal cavity obstruction, recurrent epistaxis, facial deformity, proptosis, blindness, and cranial nerve paralysis. In addition, these vascular masses bleed very easy, and its large masses may even have more than one feeding artery. Thus, it is a great challenge to establish the diagnosis and management of JNA [3, 4].

Biopsy is contraindicated in JNA cases, while surgery or angiography-embolization is the only option that can be performed to establish a diagnosis and definitive treatment in JNA cases. Therefore, careful clinical and radiological assessment will be very helpful in establishing a diagnosis and determining appropriate surgical treatment [5, 6]. Due to the limited availability of literature related to JNA in Indonesia, the author aims to explain the diagnosis and management of a case of JNA in a 13-year-old pediatric patient at Prof. Dr. I.G.N.G. Ngoerah Denpasar Hospital, Bali, Indonesia.

Case Report

The patient with the initials IMNP, a 13-year-old male, Balinese, addressed in Abiansemal, was referred from the Badung Regional Hospital on April 12, 2023, with suspected nasopharyngeal angiofibroma. The patient's main complaint is nosebleeds, which he has experienced since January 2023 on the right and left nose. Nosebleeds are said to appear intermittently and have become increasingly severe since March 2023, especially on the left nose. In the last two weeks, nosebleeds have become more frequent and numerous, approximately 3 to 4 times a week. The patient also said that his nose felt blocked, especially in the left nose, so the patient had to breathe through his mouth, and his voice sounded slurpy. Other complaints, such as headaches are sometimes felt, but double vision and ringing in one side of the ears are denied by the patient.

On physical examination, general examination was normal, aware consciousness with blood pressure 100/70 mmHg, pulse 70x/minute, respiration 30x/minute, and axillary temperature 36.5oC. On the ENT examination, there were no abnormalities in the ears and right and left external nose. On anterior rhinoscopic examination using a nasal speculum, we found masses with a smooth surface and increased vascularity in the right and left nasal cavities. The right and left mucosa are pink, while the septum and the right and left turbinates are hard to evaluate. There are no secretions. On nasoendoscopic examination, there was a mass with a smooth surface and increased vascularity in both nasal cavities. The throat examination revealed no abnormalities. The patient did not have any enlarged lymph nodes.

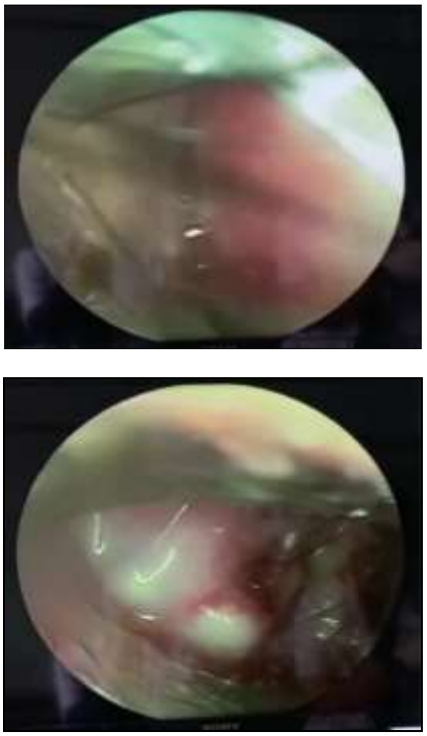

Fig 1: The nasoendoscopic picture of the patient

From the results of a CT-Scan of the nasopharynx with contrast on April 13, 2023, it was shown that there was a hypervascular solid mass in the right and left nasopharynx (predominantly left), obliterated the Rossenmuler fossa and right and left torus tubarius which extended to the left pharyngeal mucosa, right and left nasal cavities, and left masticator space. It is eroding the right and left sphenoid floor and extending to the right and left sphenoid sinus, the right and left ethmoid sinus and left frontal sinus, also eroding the medial wall of the left maxillary sinus and extending to the left maxillary sinus, as well as the ethmoid cellulae and nasal septum. This result can describe juvenile nasopharyngeal angiofibroma. It appeared the mass is getting a feeding artery from the branch of the left common carotid artery. No intracranial extension was observed.

The patient was diagnosed with suspected juvenile nasopharyngeal angiofibroma and planned to undergo a lateral rhinotomy with a left midfacial degloving and trans-palatal approach under general anaesthesia. The patient then undergoes preparations for surgery, such as chest X-rays, laboratory examinations, and anaesthesiologist consultation. There were no abnormalities found in chest and laboratory examination. From the anaesthesiologist, the patient has a physical status of ASA I. After preparation for surgery, the patient plans to undergo surgery on 10/5/2023.

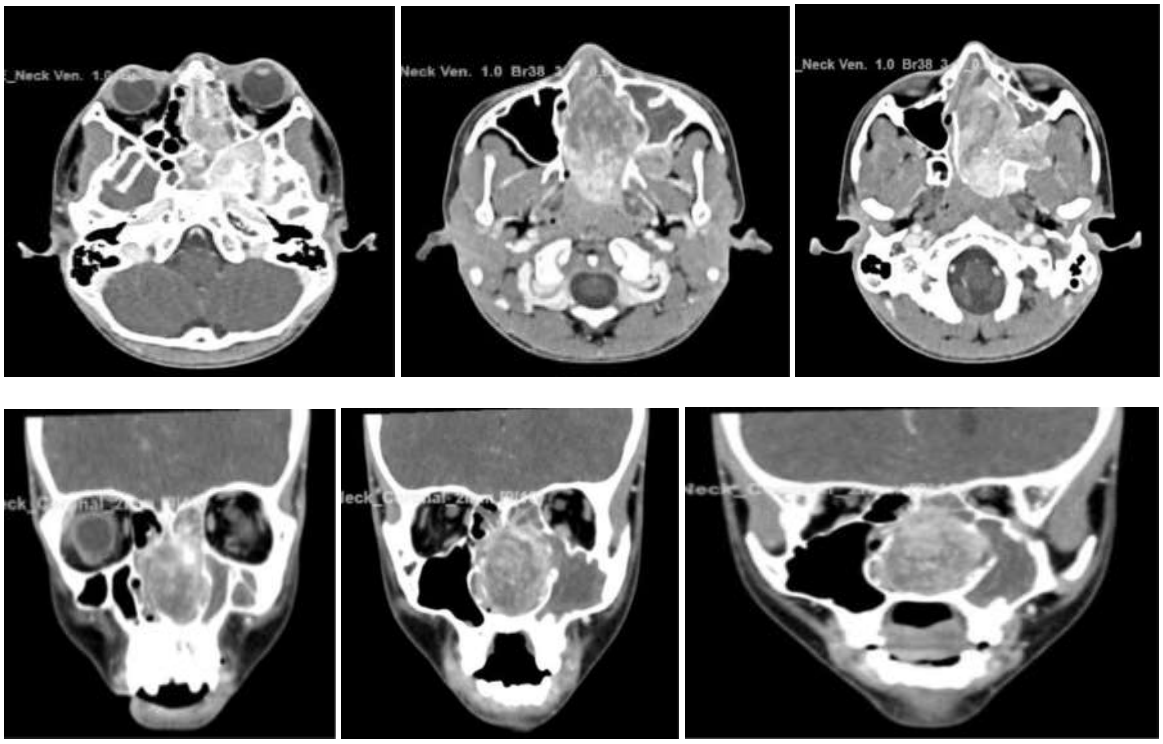

Fig 2: Axial and coronal planes of nasopharynx CT-Scan with contrast

On 10/5/2023, the patient underwent a lateral rhinotomy with a midfacial degloving left and trans-palatal approach, where a tumour mass was found in the nasopharynx intraoperatively, which extended to the left nasal cavity, maxillary sinus, ethmoid, and the left sphenoid. The incision wound is stitched, while a nasogastric tube is placed in the right nasal cavity to provide nutritional intake during the wound recovery. The procedure continued with installation of a Bellocq tampon, a sinus tampon in the left nasal cavity, and an anterior tampon in the left-right nasal cavity, which was maintained for two days.

After the surgery, the patient was treated in the PICU room to receive intensive care until his condition stabilized. He was given IVFD therapy and analgesics according to the anaesthesiologist and medication such as, ceftriaxone 2 x 1 gram i.v. and tranexamic acid 3 x 500 mg i.v. We also performed another laboratory examination and found that HB was within normal limit by 11.10 g/dL. On 11/5/2023, the patient underwent a follow-up, where his condition began to improve, the complaints such as bleeding from the nose and throat were denied, post-operative pain was still

felt but slightly reduced, soft diet via NGT, and plans to remove the tampons on the 12/5/2023.

On 12/5/2023, the patient was transferred to the room because his condition had begun to stabilize. At that time, the patient's condition was well-conscious, complaints such as bleeding from the nose and throat were denied, the post-operative pain had improved, and the patient planned to remove the tampon in the room. After tampon removal, there was no active bleeding from the nose or throat, but the NGT tube was still maintained in the right nasal cavity until the wound in the trans-palatal area improved. He was allowed to go home with the medication such as cefixime 2 x 200 mg i.o, tranexamic acid 3 x 500 mg i.o, methylprednisolone 2 x 4 mg i.o, paracetamol 3 x 500 i.o, and scheduled to return on 15/5/2023 to the ENT clinic in the oncology division for routine wound care.

On 15/5/2023, the patient came to the ENT clinic. The patient complained of a blocked nose, while the pain from the surgical wound was still felt but had improved a lot. On nasoendoscopic examination, crusts were found in the left nasal cavity, and then cleaning was carried out. The surgical wound on the hard palate showed well-maintained, clean sutures with no signs of infection or inflammation. We continue the treatment by giving cefixime 2 x 200 mg i.o, paracetamol 3 x 500 mg i.o, methylprednisolone 2 x 4 mg i.o, washing the left nose with 0.9% NaCl, and scheduled to control on 19/5/2023.

On 19/5/2023, the patient came as scheduled at the ENT outpatient clinic. His complaints of nasal congestion were improved with minimal surgical wound pain. On physical examination, we also found that the crust in the left-right nasal cavity had improved. He denied other complaints, such as bleeding from the nose and throat. The wound in the hard palate area looked clean with no signs of infection. The patient then underwent an oral eating and drinking test. We did not find any trans-palatal abnormalities or ulcers, so the NGT tube in the right nasal cavity was removed, and then continued to clean the wound in the left-right nasal cavity and washing the right and left nose with NaCl 0,9%.

On 22/5/2023, the patient came with complaints that the nasal congestion had improved, no bleeding from the nose and throat, and he was able to eat and drink. On examination using an endoscope, nasal cavity looked clear, with only a few crusts and post-operative wound residue in the nasopharynx and left nasal cavity.

The results of the anatomical pathology examination on May 22, 2023, showed a description of a nasopharyngeal angiofibroma. The patient is then educated about the disease, including the causes, symptoms and management. He then is scheduled to control until the complaint has completely improved and monitored for new complaints.

Discussion

JNA is a benign tumour that originates from the sphenopalatine foramen. Even though it is benign, this tumour can grow quickly and aggressively, causing damage to bones and surrounding structures. It is very rare, but this tumour has special predominance in the young male population. The patient in this case was a 13-year-old male patient, which is in accordance with previous literature regarding JNA [1, 7].

JNA is a vascular tumour that bleeds easily with rapid and aggressive development. It is able to develop in the sphenopalatine foramen quickly, causing pressure on the inferior and superior orbital fissures, as well as the pterygopalatine fossa. Therefore, suspicion of JNA in a young man begins with symptoms of unilateral nasal obstruction, recurrent epistaxis, and recurrent rhinorrhea. These three symptoms are the most common symptoms of JNA (75-100%). Apart from these three symptoms, JNA with extensive expansion can cause erosion of the sphenoid bone and cranial base and cause pressure on intracranial structures. Thus, JNA in advanced stages can also cause symptoms in the form of headaches, facial deformities, deafness, blindness, and even paresis of cranial nerves II, III, IV, and VI.[7,8] The suspicion of JNA, in this case, is because the patient was a 13-year-old male teenager with symptoms of recurrent nosebleeds and nasal obstruction, especially on the left side for the last three months. The patient denies any visual or auditory symptoms.

Even though the symptoms found are quite typical, the diagnosis of JNA must supported by nasoendoscopic and radiological examinations, which show the presence of a vascular mass filling the nasal cavity originating from the sphenopalatine foramen. Nasoendoscopic examination generally shows a lobulated mass behind the middle turbinate, which appears to fill the choana where the mass generally has a smooth surface and appears hypervascular.6,9,10 Anterior rhinoscopic and nasoendoscopic examination in this patient also showed a hypervascular mass with a smooth surface in both nasal cavities.

Radiological examination of the JNA functions to determine the extent of the JNA mass, which is necessary for determining the stage and planning subsequent management. Until now, a head CT scan is still the primary choice in diagnosing JNA to determining its extent, including the presence of bone erosion and invasion of intracranial structures. Head MRI examinations are less frequently performed because it is less able to provide an overview of bone invasion. However, MRI examinations may be better at determining the extent of JNA in intracranial structures.[9,10,11] Head CT-scan examinations were also performed on our patient before surgery, where the results showed a hypervascular solid mass in the right and left nasopharynx (left dominant) which obliterated the Rossenmuler fossa and right and left torus tubarius which extended to the left pharyngeal mucosa, right and left nasal cavity, left masticator space, eroded the right sphenoid floor left and extends to the left-right sphenoid sinus, right left ethmoid sinus and left frontal sinus, the medial wall of the left maxillary sinus and extends to the left maxillary sinus, and also erodes the ethmoid cellulae and nasal septum. The CT-scan image in this patient shows a typical picture of JNA, namely the Holman-Miller sign, which pushing the posterior wall of the maxillary sinus towards the anterior (anterior bowing) on the medial and lateral sides of the pterygoid. CTA or MRA examinations can also be performed in JNA cases, even though it is not routine examination. This examination aims to determine the feeding artery followed by pre-operative embolization with the aim of reducing the risk of intraoperative bleeding. Good vascular control through embolization will reduce the volume of intraoperative bleeding and the risk of recurrence. Embolization is most often performed on the external carotid artery branch which is the most common feeding artery of the JNA, but it can also be performed on other arterial branches according to the feeding artery findings

and the operator's considerations during CTA or MRA procedures. Unfortunately, in this case, no CTA or MRA examination was performed [9, 10, 11].

Staging based on clinical and radiological evaluation plays an essential role in the management of JNA. The results of clinical and radiological examinations of this case indicate that the JNA corresponds to the Fisch II, Chandler II or Radkowski IIA stage. Stage II JNA is still classified as resectable thus surgical therapy is the best modality for this case. We choose an open approach surgery, where a lateral rhinotomy was performed with a left and trans-palatal midfacial degloving approach. It was chosen because there was an expansion of the tumour inferiorly and posteriorly to the sphenoid sinus. Theoretically, surgery is still the primary therapeutic option for resectable JNA. Surgery can be performed via endoscopic or open methods, trans-palatal, trans-oral or a combination of both methods. Each approach has its advantages and depends mostly on the operator's experience and the availability of existing facilities. If the tumour is unresectable, then another treatment option is radiotherapy or hormonal therapy to reduce the size of the tumour first before performing the surgery [1, 13, 17].

Histopathological examination is generally carried out on mass obtained during surgery because biopsy is a contraindication to biopsy. It generally shows the appearance of blood vessels with a stellate or staghorn pattern with various thicknesses of blood vessel walls accompanied by stellate fibroblasts with small pyknotic nuclei to large vesicular [12, 13]. The results of histopathological examination of the tissue obtained during surgery in this case indicate the proliferation of capillary blood vessels to large dilated blood vessels, with some forming a slit-like appearance and other showing a staghorn appearance among the proliferation of connective tissue containing spindle to stellate cells with eosinophilic cytoplasm, mild pleomorphic nuclei, vesicular chromatin without sarcomatoid components in network. This result is in accordance with the histopathological description of JNA, which is a vascular mass.

JNA cases generally have a good prognosis which most cases can experience complete recovery after surgery. However, a fairly high recurrence rate has been reported, reaching 36-40%, where several factors associated with a higher recurrence rate of JNA are age <18 years when first diagnosed, large tumour size (>4 cm), and having an advanced stage [4, 20]. The patient of our case experienced significant improvement in symptoms after surgery. However, the patient was still educated regarding the disease and asked to come for control if symptoms appear again because he still has a risk of recurrence since he diagnosed at <18 years old and the tumour is >4 cm large.

Conclusion

JNA is a benign vascular tumour mass that is very rarely found with a predominance in young male patients. It can grow quickly and press on surrounding structures which can reduce the quality of life to the point of being life-threatening due to damage to the important structures that constructed the airway. Therefore, we aim that this case report will increase insight regarding JNA so that suspicion, detection, diagnosis and management of JNA can be performed as soon as possible to provide the best outcomes.

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