Preoperative Embolization in Juvenile Nasopharyngeal Angiofibroma

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Abstract
Juvenile nasopharyngeal angiofibroma (JNA) is a rare (0.05%), benign tumor with numerous vascularization, which originates from the mass formation in the sphenopalatine foramen. Incidence was mostly within age ranging from 7-19 years old, and occurrence after 25 years of age is considered rare. Preoperative embolization is a proven neuroradiological interventional endovascular technique for preoperative JNA devascularization. Male, 17 years old, presented with recurrent epistaxis in his right nose since 3 months. The patient also experienced nasal congestion, olfactory disorders, and hoarseness. History of facial trauma was denied. Neurological examinations revealed anosmia in the right nasal cavity. Funduscopy was revealed no abnormalities. Laboratory examinations findings are normal. Brain CT-Scan contrast was revealed a hypodense mass lesion within the nasopharyngeal region that has regular surface and the unclear border with contrast enhancement. The mass extend from nasopharynx area to the nasal cavity and oropharynx. Skull destruction nor nerve injury were not found, which increased the suspicion of nasopharyngeal angiofibroma. Cerebral angiography revealed a tumor blush in the nasal cavity which extends to...
sphenoid sinus and has single vascularization from a right sphenopalatine artery. Intraarterial tumor embolization was performed using polyvinyl alcohol (PVA) particles with a size of 150-250µ until no tumor blush were seen. The resection surgery of the tumor was performed 4 days after embolization with the transnasal endoscopic technique. The tumor was removed completely with minimal total blood loss during procedure (± 120cc).

Keywords: juvenile nasopharyngeal angiofibroma, embolization, tumor

Introduction

Juvenile nasopharyngeal angiofibroma (JNA) is a rare, benign well-vascularized tumor. Incidence ranged from 0.05% from tumors of head and neck. Affected age ranged from 7-19 years. The etiology of this tumor remained unknown. JNA is a very well-vascularized tumor that originates from ascending pharyngeal artery or internal maxillary artery. Preoperative embolization is a neuroradiological endovascular interventional technique that has been proven for preoperative devascularization JNA. Purpose of this preoperative embolization is to occlude or block one or more blood vessels or vascular supplies that are abnormal or malformed, which will result in tumor devascularization. Bleeding that occurred without preoperative embolization were reported about 2000cc. Since preoperative embolization is recommended for standard procedure, blood loss during surgery decreased to less than 1000cc.

Case Presentation

Male, 17 years old, presented with recurrent epistaxis in his right nose since 3 months. The patient also experienced nasal congestion, olfactory disorders, and hoarseness. History of facial trauma was denied. Neurological examinations revealed anosmia in the right nasal cavity. Funduscropy was revealed no abnormalities. Laboratory examinations findings are normal. Brain CT-Scan contrast was revealed a hypodense mass lesion within the nasopharyngeal region that has regular surface and the unclear border with contrast enhancement (figure 1). The mass extend from nasopharynx area to the nasal cavity and oropharynx. Skull destruction nor nerve injury were not found, which increased the suspicion of nasopharyngeal angiofibroma.

Figure 1. Brain CT Scan non-contrast (A) and with contrast (B) pre embolization. Mass tumor was visible on nasopharynx region with circular shape, hypodense structure, regular surface, and unclear border, with enhancement after contrast. Expansion of nasopharynx lesion to nasal and oropharynx area were also obvious. No bone nor nerve destruction were observed. Lymphatic nodes were not clear wheter there is any regional enlargement.
**Figure 2.** Cerebral angiography by injection from right internal carotid artery with anteroposterior projection (A), lateral projection (B), and right external carotid artery with anteroposterior projection (C), lateral projection (D). On figure 2A and B, normal circulation were observed without any vascular supply towards the tumor. On figure 2C and D, tumor blush were seen (white circle with dotted line) which had artery supply from right sphenopalatine artery, branch from right distal internal maxillary artery (arrow).

**Figure 3.** Intraarterial embolization procedure with Polyvynl Alcohol Particle (PVA) with size of 150-250µm. On figure 5A and B, microcatheter tip were seen proximally on right sphenopalatine artery. On angiography evaluation, there is no collateral circulation extra nor intracranially, which increase the possibility of embolization procedure to proceed. After the first PVA were administered, first angiography were done (figure 5C), and tumor blush visibility was decreasing (~50%). Figure 5D and E, tumor blush was not seen after embolization procedure. Retrograde flow from contrat was seen which indicates right sphenopalatine artery occlusion. Figure 5F angiography on right internal carotid artery, normal intracranial circulation were observed.
Figure 4. Check angiography on right internal carotid artery (above) (4A) and right external carotid artery (4B). On figure 4A, normal intracranial circulation were observed. On figure 4B, branches from external carotid artery were seen intact. However, severe vasospasm were observed on 1/3 medial of external carotid artery. CT scan with contrast post embolization (below) with saggital view (A), coronal (B), and axial (C) 7 days after embolization. Heterogeneous contrast enhancement were seen on tumor lesion that decreased >80% compared with previous imaging before embolization.

Figure 5. Tumor tissue of 4.5 x 3 x 1 cm in size, with white solid consistency, and total bleeding volume of 120cc (above). Anatomical pathology (below) with 10x magnification (A), and 40x magnification (B). Tissue were seen covered by epithelial cells, and vascular proliferation were seen subepithelially between microfibromatous stroma.
Discussion

Specific clinical manifestations of JNA is a progressive unilateral nasal obstruction (80-90%) with rhinorrhea and recurrent unilateral epistaxis (45-60%). Other clinical manifestations are a headache (25%), facial pain, unilateral otitis media, chronic rhinosinusitis, proptosis, and visual disturbance. A Headache and facial pain were resulted due to paranasal sinus obstruction. Unilateral otitis media were manifested due to disruption of eustachius tube. Expansion of tumor to sinonasal space can give a manifestation of chronic rhinosinusitis. Proptosis and visual disturbance indicate abnormalities of orbital, and swollen cheek, neurological deficit, olfactory disturbance, and otalgia can also be observed.  

In this patient, based on DSA/embolization and angiography, tumor in the nasal cavity was seen expanded to the sphenoid sinus that received single vascular supply from a right sphenopalatine artery. Anatomically, a sphenopalatine artery is a terminal branch of a maxillary artery that goes through the sphenopalatine foramen to nasal cavity, posterior part of superior meatus. Several branch and anastomosis that is related with maxillary artery like anterior temporal profunda artery, medial temporal profunda artery, foramen rotundum arteries that are related with inferolateral trunk, descending palatine artery that gives vascularization to palatum mole, descending palatine artery, sphenopalatine artery, and infraorbital artery. Temporal profunda artery sometimes was visualized as “pseudomeningeal appearance”. Foramen rotundum artery has twisted arching shape that is always going towards sellar wall, and this artery has a very important anastomosis in internal maxillary artery embolization. Management that could be done is as follows; surgery, radiation, cryotherapy, electrocoagulation, hormonal therapy, embolization, and sclerosing agent injection. Surgery is the golden standard, but huge bleeding risks are due to a highly vascularized tumor, mostly beyond 2000cc.  

Preoperative embolization is recommended as standard procedure to decrease blood loss during operation, which enables the possibility of total excision, decreasing complication, and minimalizing residual tumor. The purpose is to decrease vascular supply for the tumor, which in this case will be efficient if the embolic agent can be administered inside the tumor, and is very effective if reached by small particle like polyvinyl alcohol. Choosing particle size is to balance between safety and efficiency, as well as determining whether catheter position can be reached by direct injection of the embolic agent towards the tumor. Embolization can decrease 60-70% intraoperative bleeding. Surgery resection can be done 2-5 days after embolization. Injections with speed exceeding arterial flow can result in reflux towards the proximal arterial trunk and intracranial embolization can occur. Radiation as therapy is still debatable due to the risk of sarcomatoid transformation. Embolization in this patient was done by using polyvinyl alcohol particle with the size of 150-250 µ. According to literature, using the particle of 150-250 µ (Contour™; Boston Scientific, USA) in size can embolize small arteriole inside the tumor bed, expected to necrotize about 30-95%. Three main factor that determines the success rate of intratumoral deposition from the embolic material, which are: (a) selectivity of choosing catherization, (b) embolic material choice, and (c) no arterial spasm
on embolization. To avoid vasospasm during ECA territory, a guiding catheter was positioned only proximal from the origin of ECA or towards proximal ECA, and both (microcatheter and inserting micro guidewire) inside the ECA branch which was done in a slow manner.9

Complications that might occur during JNA embolization which were reported in the literature that can be considered serious include stroke, blindness, and paresis of cranial nerves. Stroke can occur if there is the embolic reflex of material from ECA to ICA, or when embolic materials moved from ILT branch (inferolateral trunk) and ICA towards main blood vessels. Reflex could be avoided by injecting embolic material very carefully and along with heart rate, and during systolic heart rate from arterial blood flow. Paralysis of a cranial nerve can occur when a branch of ICA or ECA gives vascularization to cranial nerves, such as blocked ILT.12 JNA is a tumor with a high recurrence rate, about 32% until as high as 40-50% in cases with basis cranii invasion.8

Conclusion
The preoperative embolization is effective to devascularization the tumor and reduce the amount of bleeding during surgery which contributes to reducing risks and complications during procedure. The prognosis of this patient, which was already mentioned above is dubia et bonam, considering that there was no involvement of the tumor towards basis cranii.

References