

Meningitis And Coma As The First Manifestation Of Juvenile Angiofibroma

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Citation

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Abstract

Juvenile Angiofibroma (JNA) is an uncommon vascular tumor of male adolescence. Although it is a benign tumor, it is locally aggressive and can erode into surrounding tissues and structures. No cause has been determined for the tumor and surgery is the treatment of choice. (1) Common symptoms are nasal obstruction, epistaxis and rhinorrhea. Meningitis at presentation has not been reported. We report a case of JNA presenting with meningitis and coma. An area of dural defect and CSF leakage was identified and repaired during the operation. Evaluation of surgical sites following surgeries in skull base should utilize surgical microscope and nasal endoscopes. This could decrease the frequency of CSF leakage and possible meningitis.

CASE REPORT

An 18-year-old man was referred to our hospital for evaluation and management of his nasal tumor. Two weeks prior to his referral he presented to another hospital with the chief complaint of headache, fever and decreased level of consciousness. Soon after his admission to that hospital he deteriorated and became comatose. Lumbar puncture suggested bacterial meningitis and culture showed *Neisseria Meningitidis*. CT scan of the brain showed a large mass at the base of skull with intra-cranial and cavernous sinus extension. For further evaluation, CT scan of the para nasal sinuses with contrast was obtained which showed a large heterogeneously enhanced lesion inside the nasal cavity and sinuses with extension to intracranial, intraorbital, infratemporal fossa and left cheek. The posterior wall of the left maxillary sinus was pushed forward (Holman Miller's sign). Cavernous sinus and anterior cranial fossa were involved with the tumor, which classified tumor as stage 4a according to Andrew's staging. With these findings patient was referred to us for further management. Upon more detailed history patient reported nasal obstruction, rhinorrhea and epistaxis few months prior to his meningitis. Physical exam revealed a grayish-red massive mass in posterior nares and left side of the nasopharynx with intact mucosa. Mild proptosis of the right eye was seen. There was a firm mass with wormy sensation felt in his left cheek. Visual acuity was 20/20 bilaterally. Rest of the physical exam was unremarkable. No spontaneous or valsalva induced CSF leakage was found during physical examination. All

hematology and chemistry tests were within normal limits.

With the diagnosis of Juvenile Angiofibroma surgical resection was recommended. After obtaining informed consent, patient was scheduled for surgery. Under general hypotensive anesthesia and via a transpalatal and transantral approach tumor was identified and resected completely. (Fig 1) After the resection the tumor site and area was examined carefully using surgical microscope and 4-mm, 0 and 30degree nasal endoscopes. An area of herniation of the dura with some defect was found. The defective area was about 8X8 mm and showed mild intermittent CSF leakage. Elevating the dural herniation precipitated CSF leakage. Dural defect was reconstructed using fascia lata muscle graft, surgicel and posterior and anterior nasal packing. During the operation patient was transfused with 4 units of RBC. He was started on 2 grams of ceftriaxone twice/day. On first post-operative day, he ran a temperature of 38.5. Physical examination was unremarkable; there were no meningeal signs. Lumbar puncture was done which was normal. Chest X-ray and other Laboratory tests were all within normal limits. CT scan showed no hematoma or mass effect. Treatment with ceftriaxone and vancomycin was continued. Patient spiked fever for two more days, but temperature dropped after removing the nasal packing on day 4. There was no CSF leakage or other complication and he was discharged from the hospital seven days after the surgery. We have followed the patient closely since the surgery 8 months ago. Physical exam, CT scan and nasal

endoscopy have been all normal.

DISCUSSION

Juvenile Angiofibroma (JNA) is a benign tumor of young men. The triad of nasal obstruction, recurrent epistaxis and nasopharyngeal mass are highly suggestive of this tumor. Although the tumor is histologically benign, it can be aggressive and destructive. JNA accounts for 0.5% of all head and neck tumors. Incidence is one case per 5,000 to 6,000 ENT patients. The cause has not been determined yet. The most accepted theory is that JNA originates from sex steroid hormone stimulated hamartomatous tissue located in the turbinate cartilage. Another proposed theory includes tumor originating from embryonal chondrocartilage of the occipital plate. (2) Intracranial extension of JNA is either through the sella medial to the carotid artery and lateral to pituitary gland or via erosion of the greater wing of sphenoid through the middle cranial fossa anterior to the foramen lacerum and lateral to cavernous sinus and carotid artery. (3) Usually signs and symptoms present for 6 months prior to diagnosis. CT scan of the head and facial bones with contrast confirms the clinical diagnosis and shows the extension of the tumor. Different staging systems have been used for staging JNA. Andrew's staging system is shown in table 1. (4) Modalities used for treatment of JNA include surgery, hormonal therapy, radiation treatment and chemotherapy. Surgery is usually recommended for stages 1 to 4a. Radiation with or without surgery is usually done for stage 4b. External beam radiation is most often reserved for intracranial extension and unresectable tumor or recurrence of the tumor. Different approaches have been used for surgical resection of the tumor. Extension of the tumor and the surgeon's expertise guide the choice of any particular approach.

Although there are reports of meningitis after surgery for JNA, we could not find any report of JNA that presented initially with meningitis and coma. CSF leakage and meningitis are most often caused by invasion of malignant tumors. In about 20% of all major cranial base surgeries, CSF leakage can occur. (5) There are some reports of meningitis following the surgeries. In our case the causative organism of the meningitis was Neisseria meningitidis, which is the most common cause of bacterial meningitis in children and second most common cause in adults. Since there was no CSF leakage found in physical exam prior to the surgery, we cannot be sure that the route of infection was laceration in dura. But because we were suspecting the possibility of a dural defect, surgical site was fully searched

with surgical microscope and nasal endoscopes. This search showed the herniated dura, which was leaking CSF around it while elevated. We believe that this patient had had intermittent CSF leakage and because it was minimal he never noticed it and we could not see it during our physical examination. This case emphasizes the importance of thorough evaluation of the surgical sites after skull base surgeries. By using surgical microscope and especially nasal endoscopes all areas including areas that are difficult to visualize could be easily seen and any dural defect could be repaired. This procedure may help to decrease the prevalence of meningitis and CSF leakage after skull base surgeries.

Figure 1

Table 1: Andrew's classification of JNA

<ul style="list-style-type: none">o I - Tumor limited to the nasal cavityo II - Into pterygopalatine fossa or maxillary, sphenoid, or ethmoid sinuseso IIIa - Into orbital or infratemporal fossa without intracranial involvemento IIIb - Stage IIIa with extradural intracranial (parasellar) involvemento IVa - Intradural without cavernous sinus, pituitary, or optic chiasm involvemento IVb - Involvement of the cavernous sinus, pituitary, or optic chiasm
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Figure 2

Figure1



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