Abstract

Neurofibromas are benign peripheral nerve sheath tumors. They arise from schwann cells and fibroblasts of the connective tissue of the endothelium. Of the 25% to 45% neurofibromas that arise in the head and neck region, only 4% involve the nasal cavity and paranasal sinuses. Here we report one such case in a 18 years old male who presented to us with complaints of swelling in his left cheek since a year ago which often felt to shrink and then enlarge again and also complains of several other symptoms like watering of the left eye frequently, nasal obstruction, and also crooked tooth. The mass was histologically confirmed to be a neurofibroma, but Computer tomography and panoramic image leads it to dentigerous cyst.

Keywords: sinonasal mass, neurofibroma,

Introduction

Neurofibroma consists of the word "neuro" which means nerves and "fibroma" is a tumor that occurs in connective tissue (fibrous). Neurofibroma are a benign tumor arising in intimate association with a peripheral nerve trunk, developing from Schwann cells, perineurites, and blended with fibroblastic cells. Neurofibroma are growths of Schwann cells (producing nerve sheaths or myelin) and other cells that surround and support peripheral nerves (peripheral nerves, nerves outside the brain and spinal cord)). (1) Neurofibroma is divided into solitary mass and multiple mass which include neurofibromatosis I and II. Schwann cells and perineural cells derived from neuroectoderm are thought to be the origin of all these neoplasms. In the area of the nose and paranasal sinuses, neurofibroma arises from the first and second division of the trigeminal nerve and from autonomic plexuses. (2,3)

Although they may occur in any part of the body, the occurrence within the nasal cavity and paranasal sinuses are uncommon. Schwannomas (about two-thirds) are more frequent in the nasal cavity and paranasal sinuses than neurofibromas (one-third). The nasal cavity alone is most commonly affected, followed by maxillary sinus, or mixed sites. (4)
Case Report

An 18 years old male presented to us at the polyclinic IK-THT-KL with a history of swelling of his left cheek and a mass protruding from his left maxillary sinus area. He gave us a history of the mass being present in his cheek since a year ago and was slowly growing in size. He said that this mass is suddenly enlarge and shrink without any triggered. Since the mass was growing in size and he does not comfort about it, he decided to go to RSUD Cut Meutia to check the real diagnosis about his condition.

He also said that he has nasal obstruction and his left eyes often watering frequently. He also realized that his left incisor was crooked due to his swelling. He did not give a history of any other abnormal growth or lumps being present over his body or extremities.

Figure 1 sinonasal mass on left cheek's patient

On physical examination, it looks like a mass filling the front of the left nasal cavity pressing the left inferior turbinate medially, no bleeding is found, the color is the same as around. On the oral examination, there was no protrusion of the palate, brittle teeth and masses in the oropharynx. There was a mass pressing on the left front gum with a diameter of 1 cm x 2 cm x 0.5 cm. There were no palpable neck nodes. On examination her ears and throat were normal.
The x-ray image showed a mass in the left nasal cavity and extending up to the maxillary sinus causing pressure symptoms on the septum and pushing it to the right. Darkness in part of the left nasal cavity up to the maxillary sinus, the sinonasal mass extends to the maxillary sinus. The left maxillary sinus was opaque.

Accordingly to the patient was evaluated for surgical fitness and the mass was biopsy under anesthesia. There was not much hemorrhage during surgery and the patient tolerated the procedure well and post operative period was uneventful.

The histopathological examination confirmed the presence of spindle nuclear cells, sufficient cytoplasmic, fine chromatin, inflammation of lymphocytes, histiocytes and plasma cells in other areas which confirmed the diagnosis of neurofibroma. There are no specific processes and signs of malignancy in this preparation.
By the CT scan image coronal part and axial there is a cystic lesion with indistinct borders, regular edges in the left maxillary region with a size of 5.56 x 5.02 x 3.59 which with contrast administration did not show abnormal contrast enhancement, the lesion appears to fill the left maxillary sinus cavity there was no hypodense/hyperdense lesion in the brain parenchyma which did not show abnormal contrast enhancement with contrast administration, normal sulci and gyri, normal ventricular and cystic systems, normal pons and cerebellum, no abnormal calcifications, no midline deviation, orbit and mastoid right and left no abnormalities, normal calvaria. And the conclusion for this CT scan image leads to dentigerous cyst and there is no infarction or mass seen in the brain parenchyma. By this finding we decided to confirm the diagnosis clearly with panoramic photo whether it still leads to dentigerous cyst or not.

On this panoramic photo showed there is no lucency and no filling on the crown of the teeth, no visible part of the teeth is in the alveolar process, no destruction of the
mandibular or maxillary bones was seen, and there is no missing teeth. It only showed a dental impaction in 48 teeth (messioangular).

**Discussion**

Neurofibroma is a benign tumor of the connective tissue mainly involving the endoneurium of peripheral nerves. Peripheral nerve sheath mass are divided into schwannoma, neurofibroma, and neurogenic sarcoma. They rarely arise in the area of the nose and paranasal sinuses, especially neurofibroma. Neurofibroma can occurs in the head and neck about 25-45% but only 4% are found within the nasal cavity. (5)

Neurofibroma in the area of the nose and paranasal sinuses, as other neoplasms in this area, leads to non-specific symptoms including nasal obstruction, epistaxis, cheek swelling, facial pain and proptosis. Thus the histological examination is essential to the diagnosis. We found Clinical symptomatology in this case is any process occupying the nasal and sinus cavities. It comprises nasal obstruction, which in the present case could became bilateral due to the mass that push into the right side of nasal and leads to septum deviation. (5) Neurofibroma seems to develop from the 1st and 2nd branches of the trigeminal nerve innervating various nasal and sinus cavity structures, notably including turbinates and maxillary sinus. (6) This condition could disturb the function of trigeminal nerve as in this patient there is a watering of his left eyes frequently without any triggered. 

Macroscopically the mass appears as a gray white color mass with a firm consistency. Histologically, the mass consists of proliferation of spindle nuclear cells, fine chromatin, sufficient cytoplasm, in other areas the distribution of inflammation of lymphocytes, histiocytes and plasma cells was also seen.

The treatment of neurofibroma is complete resection of the mass because neurofibromas may infiltrate extensively. Other treatments are not effective. In more extensive cases, the extensive operation should be performed. Recurrence is rare, but we should keep in mind that both mass may have the potential of malignant transformation. The malignant transformation is reported to be at the rate of 10%. (7)

**Conclusion**

Neurofibroma are benign peripheral nerve sheath tumors that arise from schwann cells and fibroblasts of the connective tissue of the endothelium. Neurofibroma in the nasal cavity and paranasal sinuses are very uncommon. In this case there are several symptoms
performed like swelling on the cheek and also nasal obstruction. By the pathological anatomy it leads to neurofibroma.

References