Large Neurofibroma of Trunk

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Abstract Neurofibroma is a benign tumor of cutaneous nerves. These are benign tumors which may have a varied presentations ranging from a cosmetic problem to a spinal tumor which may lead to neural complications. We here by present a case where the patient has become an appendage of the tumor and its the mass of the tumor that has handicapped the patient.

Keywords Neurofibroma trunk

Background

Plexiform neurofibroma is an uncommon variety of neurofibromatosis, usually associated with trigeminal nerve. It has also been reported in retroperitoneum, paraspinal and mediastinal area. A single case of anterior abdominal wall plexiform neurofibroma has been documented by Aloï et al [1].

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We report a rare case of segmental plexiform neurofibroma over the anterior and posterior abdominal wall.

Our Case

A 52 year old male presented to us large fleshy lesion hanging from back and side of abdominal wall since age of 5 years. Initially they were short which gradually enlarged to acquire the present size. Due to the large size his activities became restricted and gradually he became incapable of even carrying out his daily activities.

On examination he had large neurofibromas hanging from anterior abdominal wall and back.

He had a short stature with thoracolumbar kyphoscoliosis. There were cafe-au-lait spots all over the body.

An irregular pendulous mass, 30 × 70 cms on left lumbar region and 30 × 45 cm on right lumbar region was hanging superficial to the anterior abdominal muscles. It was non tender, firm and pedunculated. Thickened nerves could be palpated. The sensations were normal.

Systemic examination was normal with no neurological deficit. Plain radiograph of the chest did not reveal any mediastinal widening and radiograph of the skull did not show any widening of internal acoustic meatus. Computed tomography of the abdomen did not reveal any intra-abdominal abnormality.

Discussion

Plexiform neurofibroma is neurofibromatosis is a generalized overgrowth of neural tissue occurring in subcutaneous tissue [2–5]. Its cellular lineage is uncertain, and may derive from Schwann cells, other perineural cell lines, or fibroblasts. This

condition is autosomally dominant with variable penetration and presents as multiple nodules of various sizes, which are firm and nontender associated with café-au lait spots. The condition is usually associated with spinal deformities in form of kyphosis or scoliosis.

It is usually present along distribution of trigeminal nerve but can have segmental presentation [1, 6]. Segmental presentation are usually asymptomatic and produce cosmetic disfigurement. They may cause functional impairment due to the bulk of their size. They may have neuro deficit incases of spinal neurofibroma.

Diagnosis id made clinically. The National Institute of Health (NIH) has created specific criteria for the diagnosis of NF-1. Two of these seven “Cardinal Clinical Features” are required for positive diagnosis [7]. Ultrasonography may show hypoechogenicity. CT scan and MRI are indicated incases of neurofibroma of spinal cord [8–10].

It is a benign lesion but malignant changes can occur. MRI may be helpful in diagnosis of malignant change. 67 Ga citrate scintigraphy has been advised in-patients of neurofibromatosis with suspected malignant change [11] as only malignant cells have been found to take up 67 Ga.

Treatment is indicated in cases where the impairment is caused due to bulk of tumor. In symptomatic neurofibroma of spinal canal decompression is to be carried out. Thorough search is to be made for retroperitoneal medistinal paraspinal neurofibroma as they notorious to undergo malignant changes. Excision with skin grafting is done cosmetically important areas. Patients are to be given genetic counseling.

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