

Vagal Cervicomedial Neurofibroma in Neurofibromatosis Type 1 mimicking Pulmonary Tuberculosis in Infancy: A Case Report

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Summary

Odebode TO, Ologe FE, Adeoye OPO, Afolayan EAO, Aluko AA, Odelowo EOO. Vagal Cervicomedial Neurofibroma in Neurofibromatosis Type 1 mimicking Pulmonary Tuberculosis: A Case Report. *Nigerian Journal of Paediatrics* 2005;32:140. Mediastinal neurofibroma, a rare complication of Neurofibromatosis type 1, (NF1), rarely mimics tuberculosis. We report the case of a 12-year old boy who presented at a health establishment during infancy with clinical and roentgenographic features suggestive of pulmonary tuberculosis for which he received antituberculosis drug therapy. This time, he presented with hoarseness, left ptosis, posterior neck gibbus and dermal nodules. Radiograph and computed tomography (CT) of the chest showed a left cervicomedial tumour extending to the right superior mediastinum. At thoracotomy, a globular tumour measuring 10x7x6 cm and arising from the left vagus nerve was resected with a segment of the nerve. The cervical extension and a rim encroaching on the left brachial plexus were spared to minimize post-operative morbidity. The histopathological diagnosis was neurofibroma. Although neurofibroma originating from the vagus nerve has been reported before, this, to our knowledge, is the first reported case from the middle-belt of Nigeria. Since vagal neurofibroma can manifest earlier than cutaneous neurofibroma in patients with NF1 and mimic other chest lesions, clinicians should endeavour to investigate suspicious infantile chest lesions with other modalities in addition to chest radiography.

Keywords: neurogenic, mediastinal, vagus nerve, tumour

Introduction

NEUROFIBROMATOSIS type 1(NF1) is an autosomal dominant multi-system genetic disorder with an estimated prevalence of 1 in 4000 live births.¹ The

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criteria for the diagnosis of dermal, but not mediastinal neurofibromas were established in 1988.² Approximately 30 to 45 percent of mediastinal neurofibromas occur in individuals with NF1 and could arise from peripheral nerves or from autonomic ganglia.³ Most of these tumours are asymptomatic, being detected incidentally during radiological investigations.³ However, they present occasionally with compressive or neurological symptoms especially when they become malignant, usually in patients with long-standing NF1.³ Therefore, resection is advisable once detected.

Bilateral cervicomedial neurofibroma of vagal nerve origin in patients with NF1 has been reported before.⁴ However, in this communication, we report a unilateral form originating from the left vagus nerve in a child with NF1, who had earlier received treatment for suspected pulmonary tuberculosis.

Case report

A 12-year-old boy presented with left ptosis, posterior neck gibbus and hoarseness of the voice of five years' duration. He had a few unrecognized *café-au-lait* macules from birth but developed dermal nodules (neurofibromas) only three years before presentation. He had presented elsewhere during infancy with chronic cough, fever, night sweats and weight loss. The chest radiograph, which showed a left upper lobe opacity led to a diagnosis of pulmonary tuberculosis for which he received a six-month course of anti-tuberculosis drugs with clinical improvement, although the



Fig. 1 Twelve-year-old NF1 patient with a short neck, cervical kyphosis (arrowed) and occipital bossing.

pulmonary lesion had persisted.

Clinical examination revealed a boy with a short stature (height, 1.3m), a short webbed neck, cervical kyphosis (Fig. 1), widespread truncal *café-au-lait* spots and three subcutaneous neurofibromas. On slit lamp examination, there were 16 iris Lisch nodules in both eyes but the visual acuity was normal (6/6). He had a left ptosis, anhydrosis and enophthalmus (Horner's syndrome) as well as hypertelorism, down-slanting palpebral fissures, malar hypoplasia and low-set ears (Noonan's phenotype). There was no behavioral abnormality, neither was there cognitive deficit or specific learning disability. The left vocal cord was paralyzed on indirect laryngoscopy. Lateral radiograph of the neck revealed erosion and deformity of vertebral bodies and their posterior elements with cervical kyphosis and elongated spinous processes. Antero-posterior chest radiographs revealed soft tissue cervicomediastinal shadows with



Fig. 2: Posteroanterior chest radiograph of the patient showing a left superior mediastinal shadow.

pressure erosion and deformity of ribs (Fig. 2). A contrast-enhanced CT of the neck (Fig. 3) and chest (Fig. 4) showed a left postero-superior mediastinal tumour extending anteriorly into the lower cervical region. The electrocardiogram and brain scan were normal.

At left posterolateral thoracotomy, a firm globular tumour measuring 10 x 7 x 6 cm and arising

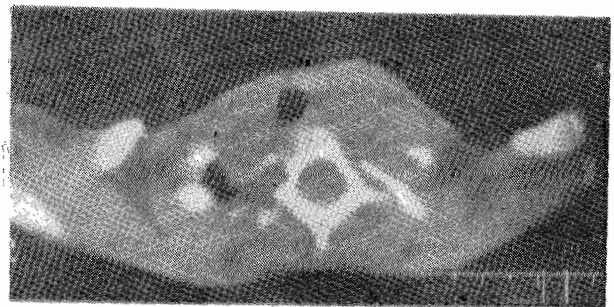


Fig. 3: Contrast-enhanced neck CT showing left cervical tumour extension, tracheal deviation and pressure erosion with deformity of adjacent vertebral bodies and posterior elements.

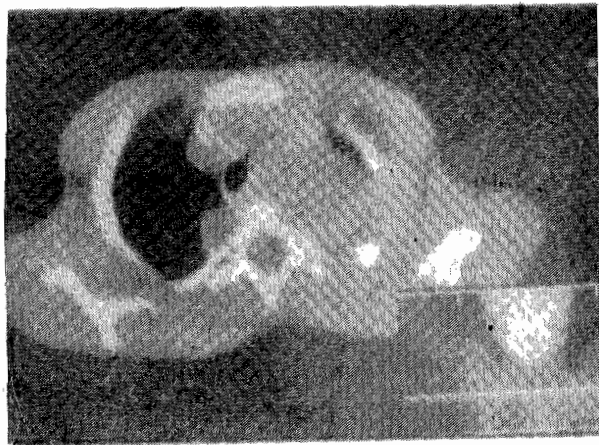


Fig. 4: Contrast-enhanced chest CT of the same patient showing a left posterior mediastinal tumour extending to the right mediastinum and displacing adjacent trachea and lung.

from the left vagal trunk (Fig.5) was resected with a segment of the vagus nerve, sparing its cervical extension and a rim abutting on the inferior cord of the left brachial plexus. Involvement of this plexus was suspected when dissection of the supero-lateral mediastinal component elicited vigorous contraction of the left upper limb muscles. The histopathological diagnosis of the tumour was benign neurofibroma characterized by wavy rows of spindle cells without



Fig. 5: Intra-operative photograph of the spherical tumour (arrow) arising from the vagus nerve.

cellular atypia (Fig. 7). The patient has enjoyed a post-operative course of one and a half years without clinical deterioration or radiological evidence of recurrence.

Discussion

In the series reported by Zhang, Ping & Bai, neurofibroma accounted for 27.2 percent of 125 surgically treated primary neurogenic mediastinal tumours;⁵ 62 percent of these patients had no subjective symptoms. It is thus possible for an attending physician as apparently happened in the early stages of our case, to misinterpret the clinical features. A high level of awareness of NF1 and its complications is required for instituting appropriate radiological investigations and making a prompt diagnosis.

The current case presented in a rather unique manner by combining dermal neurofibromas, which he developed at age nine, cervicomediastinal neurofibromas, secondary cervical kyphosis and Horner's syndrome with a Noonan's phenotype without cognitive impairment. Colley *et al*⁶ had earlier reported Noonan's phenotype in 6.4 percent of their patients with NF1. The left vagus nerve is more frequently affected by a neurofibroma than the right.⁴ This is probably due to the fact that the proximal intra-thoracic vagal trunk is thicker on the left than the right side.⁴ Most reported cases of vagal tumours are unilateral, the bilateral forms being rare.⁴

Mediastinal neurofibroma is associated with an increased risk of malignancy especially in patients with NF1.³ It is best, therefore, to resect them as early as they are detected, not only to confirm the nature of the lesion, but also to prevent further growth and compression of adjacent structures. For benign tumours, this is curative. Tumour resection was partial in our patient to avoid injury to the brachial plexus, and thus minimize postoperative morbidity. For the same reason, only a segment of the vagus nerve was resected. Tumour resection could be performed with vagus nerve amputation or by intra-capsular excision without nerve amputation.⁷ However, it has been argued that it may be difficult to partially extirpate a vagal tumour while safely preserving the nerve, since neurofibromas contain all nerve elements including axons, sheath cells, and connective tissue.^{3,4} Traditionally, resection has been performed by a standard posterolateral thoracotomy,⁸ as in this case. More recently however, such tumours have been removed thoracoscopically.⁸ A cystic lymphangioma of the heart mimicking a mediastinal tumour has been reported,⁹ but this is the first report of a cervicomediastinal tumour mimicking pulmonary tuberculosis.

We wish to emphasize the fact that vagal neurofibroma could manifest earlier than cutaneous

neurofibroma in patients with NF1, and mimic other chest lesions. Clinicians should therefore, widen the investigation of suspicious infantile chest lesions beyond plain chest radiography.

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