Ancient Cervical Vagal Schwannoma: A Diagnostic Challenge

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Sir,

Nerve sheath tumors arising from the cervical vagus nerve are extremely rare and difficult to diagnose. We report a case of a 33-year-old male who presented to us with a two year history of an asymptomatic left sided cervical swelling. Clinical examination revealed a firm, well circumscribed 14x6cm neck mass with minimal transverse mobility along the nodal levels II to IV. Clinical examination of the rest of the neck, the upper aero digestive tract and the cranial nerves were unremarkable. A computerized tomography scan revealed an irregular hetero dense well enhancing mass with multiple calcifications in the left neck extending from the level of the C2 vertebra to the level of the D2 vertebral level measuring 14×5.9×4.6cm, displacing the carotid artery anteriorly and severely compressing the left internal jugular vein. A digital subtraction angiography revealed the mass to be mainly supplied from one of the branches of the external carotid artery (Fig. 1a, b). Fine needle aspiration cytology of the mass repeated twice was inconclusive. The patient was therefore taken up for a surgical exploration with a provisional clinical diagnosis of a soft tissue tumor.

Intra operatively an encapsulated yellowish white mass was seen in close proximity to the vagus nerve (Fig. 2a, b). The tumor was meticulously separated from the trunk of the vagus nerve, the carotid artery and was removed en-bloc

along with the internal jugular vein using micro surgical dissection techniques. The patient made an uneventful clinical and neurological recovery.

The post operative histopathology revealed a 15x8x5cm encapsulated mass, the cut surface of which appeared yellowish white with myxoid areas. Microscopic examination revealed a tumor composed of spindle cells with vesicular nuclei surrounded by a markedly hyalinized stroma. Most of the areas show marked myxoid degeneration and hyalinization (Antoni B) and focal cellular spindle cell areas with dark staining nuclei. (Antoni A) Scattered calcifications are seen

Scattered calcific spherules, hyalinized blood vessels and lymphoid aggregates were also seen, suggesting a diagnosis of an Ancient Schwannoma (Fig. 3).

Schwannoma is a rare benign neural tumor, arising from the neural sheath schwann cells of the peripheral, cranial or autonomic nerves. Ackerman and Taylor in 1951 [1] coined the term “ancient” schwannoma to describe a long standing degenerative schwannoma that presented with wide areas of hyalinized matrix. Despite the degenerative changes ancient schwannomas are believed to have a clinical behavior similar to that of a conventional schwannoma. Schwannoma originating from the cervical vagus nerve is an extremely rare neoplasm. They usually occur between the third and fifth decades of life, with no definite sex predilection.

The pre-operative diagnosis of schwannoma is a clinical challenge and is aided by imaging. MRI scan with gadolinium contrast is said to be the preferred technique of choice for imaging. The imaging findings are also useful in providing a clue as to the nerve of origin of a cervical schwannoma. The vagal schwannomas displaces and separates the internal jugular vein and the carotid artery, whereas schwannomas from the cervical sympathetic chain displaces both the carotid artery and internal jugular vein without separating them [2].

Complete surgical excision is the treatment of choice of cervical vagal schwannomas. Transecting and re-anastomosing

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Fig. 1 **a** A computerized tomography scan revealed an irregular hetero dense well enhancing mass with multiple calcifications in the left neck extending from the level of the C2 vertebra to the level of the D2 vertebral level measuring $14 \times 5.9 \times 4.6$ cm, displacing the carotid artery

anteriorly and severely compressing the left internal jugular vein. **b** A digital subtraction angiography revealed the mass to be mainly supplied from one of the branches of the external carotid artery

Fig. 2 **a** Intra operative photograph showing an encapsulated yellowish white mass was seen in close proximity to the vagus nerve. **b** The post operative specimen photograph

Fig. 3 **a, b** H&EX10: Most of the areas show marked myxoid degeneration and hyalinization (Antoni B) marked as B and focal cellular spindle cell areas with dark staining nuclei (Antoni A) marked as A. Scattered calcifications are seen (C). Few blood vessels are also seen surrounded by hyalinized stroma

the vagus nerve invariably results in significant morbidity, hence attempts at nerve preservation should be made whenever possible considering the benign nature of a majority of the cervical vagal schwannomas [3, 4]. Some series have reported the incidence of post operative vocal cord paralysis as high as 85%, citing difficulty in separately identifying the trunk of the vagus nerve from the tumor [5]. This was however not the case in a few other series [3, 4] as also our patient, who had an uneventful neurological recovery.

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