Role of FNAC in the Diagnosis of Carotid Body Paraganglioma

Case Report

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ABSTRACT

This is a case of carotid body paraganglioma diagnosed on fine needle aspiration (FNA) cytology prior to other diagnostic work up. A lateral neck mass with a long duration, the hemorrhagic nature of the aspirate and the cytological features; some of which were reminiscent of an endocrine neoplasm are the factors that helped in suggesting the possibility of paraganglioma which was later confirmed in histopathology. We report this case to highlight two points: 1) the importance of FNAC in diagnosing such lesions in clinically obscure cases. 2) In FNAC of neck lesions, if paraganglioma is suspected, preparation of a cell block along with immunohistochemistry in cases wherever possible will prove useful for the definitive diagnosis of paraganglioma.

KEY WORDS: carotid body paraganglioma, cell block preparation, clinical history, fine needle aspiration, histopathology

INTRODUCTION:

FNAC has been firmly established as the preliminary diagnostic test for the evaluation of various neck lesions. However, there has been a controversy regarding its utility in the diagnosis of carotid body paraganglioma. Hemorrhage within the tumor and damage to the carotid artery are the anticipated complications which led to the objection of this test for evaluating such lesions.[1] But in clinically obscure cases, FNAC is the preliminary investigation sought for and the cytopathologist is called upon for diagnosis. The case presented here unravels the difficulties related to the diagnosis of carotid body paragangliomas in cytology, the importance of clinicopathologic correlation and some practical suggestions which would prove useful to arrive at a definitive diagnosis.

CASE REPORT:

A 52 year old male presented with a left sided neck mass which had been progressively increasing in size over the last 6 years. On examination, the mass was of size 3x2 cm and was firm in consistency. The patient had been treated in another centre as a case of lymphadenitis. He had a CT report which showed an enhancing left sided neck lesion. FNA of the mass was performed using a 23 gauge needle attached to a 5ml syringe. The initial aspirate yielded blood only. The procedure was repeated and the material obtained was spread on glass slides. Wet smears fixed in 95% isopropyl alcohol were stained by papaniclaou method. Dry smears were also prepared and stained using May Grunwald Giemsa (MGG) stain. The cytology smears were cellular with cells arranged in clusters and sheets in a hemorrhagic background. The possibility of paraganglioma was suggested in view of the clinical history, nature of aspirate and cytology findings.

All preliminary investigations were done and the patient underwent surgery. Intraoperatively, a 5x4 cm pulsatile mass was seen on the left side of the neck extending from the lower end of the mandible to the lower one-third of the sternocleidomastoid muscle. The mass was resected and sent for histopathological examination; and the biopsy report confirmed the diagnosis.

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Pathologic Findings were as follows:

FNAC: The cytology smears showed cells in clusters, sheets and follicular pattern in a hemorrhagic background (Figure1A).The cells have round nuclei and display marked anisokaryosis with moderate cytoplasm showing scattered red granules (Figure 1 B). Cells with round to spindle nuclei and fine granular chromatin seen (Figure 1C). No mitosis or necrosis was seen.

Histopathology: The biopsy specimen showed a neoplasm composed of cells arranged in the classic zellballen pattern (Figure 2A) and nests interspersed by hyalinised blood vessels. Cells had moderate amount of eosinophilic granular cytoplasm and showed marked anisokaryosis with stippled chromatin (Figure 2B). Mitosis was not evident.

DISCUSSION:

The differential diagnosis of lateral neck masses vary from benign to malignant lesions. These include tuberculous lymphadenitis, branchial cleft cyst, metastatic carcinoma, paraganglioma, schwannoma and lymphoma.[2, 3] Since FNAC is a simple, noninvasive procedure which provides a valuable clue regarding further management, in most instances the patient is sent to the cytopathologist for a preliminary diagnosis.

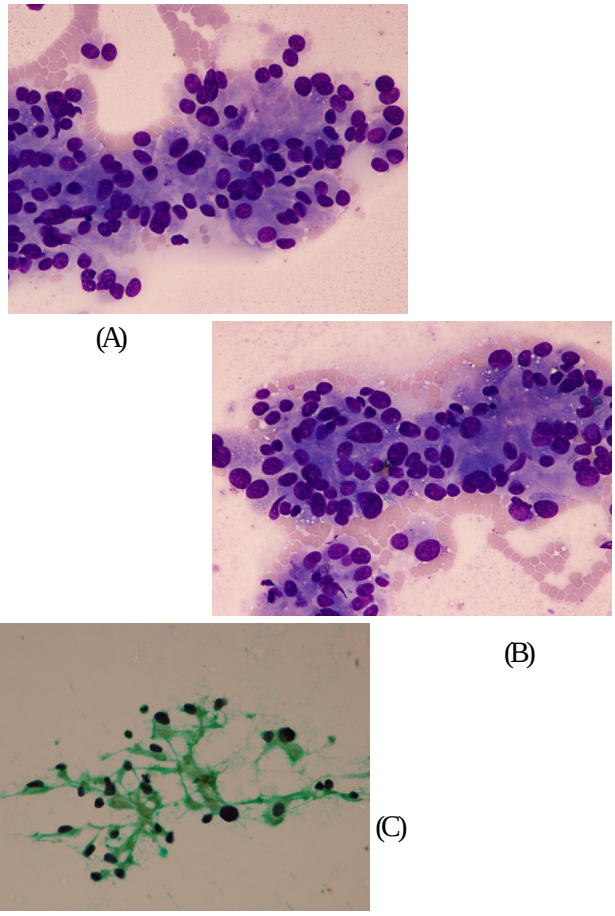

Figure 1: A-C FNA cytology A. Tumor cells in sheets and vague follicular pattern (MGG ×400). B. Cluster of tumor cells showing indistinct cell borders, moderate to abundant cytoplasm and anisokaryosis (MGG ×400). C. Cells with round to spindle nuclei and fine granular chromatin are seen (PAP×400)

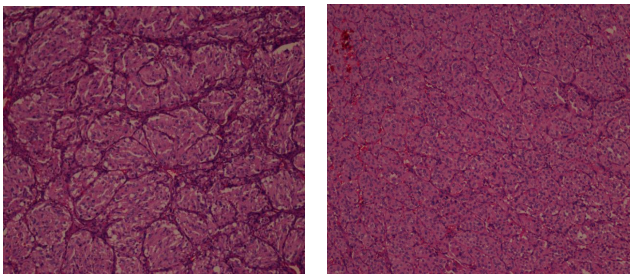

Figure 2: A and B. Histopathology specimen A. Neoplastic cells in the classic zellballen pattern. (hematoxylin-eosin stain x 10). B. Cell nests displaying marked anisokaryosis (hematoxylin-eosin stain x 10).

Paraganglia are collections of specialized neural crest cells arising in association with the autonomic ganglia. Tumors arising from these cells are called Paragangliomas and their nomenclature depends on the anatomic location of the lesion.[4] Carotid body tumor is the most common extra adrenal paraganglioma and it is situated at or around the carotid artery bifurcation**.** The usual clinical presentation is that of a painless, slowly enlarging cervical mass and the average age at diagnosis is usually the fifth decade of life. Preoperative diagnosis is possible in clinically suspected cases with the help of radiological investigations like CT scan, MRI, ultrasound and selective digital subtraction angiography. These tests also help in delineating tumor location and defining blood supply.[6] In cases where the clinical picture is obscure, the help of the cytopathologist is sought for to arrive at an initial diagnosis. In our case, the long duration of the swelling and the hemorrhagic nature of the aspirate was a clue that helped us to include paraganglioma in the differential diagnosis.

On review of literature**,** the reported cases of carotid body paraganglioma diagnosed in FNAC showed more or less similar features. The tumor cells had moderate cytoplasm and round to oval nuclei showing anisokaryosis. The nuclear chromatin was evenly distributed with inconspicuous nucleoli. In most of the cases, mitosis was either absent or very low. The hemorrhagic background was a common feature in all the cases. In three reports, the presence of prominent intranuclear vacuoles was mentioned bringing thyroid carcinoma into the differential diagnosis.[5] Features like rosette formation and cell

Table 1: FNA Diagnosis in reported cases of Carotid body Paragangliomas.

		FNAC Diagnosis				
Author series	Number of cases	Initially as paraganglioma	Revised to paraganglioma	Other diagnosis	Diagnosis not presented	Unsatisfactory specimen
Engzell et al.[1]	13	7		2 thyroid carcinoma 2 neurofibrosarcoma 1 neurofibroma		1
Hood et al.[4]	1			1 atypical epithelial lesion		
Jacobs et al.[5]	1			1 thyroid neoplasm		
Kapoor et al.[2]	1	1				
Gonzalez et al.[6]	2				2	
Chen et al.[7]	1	1				
Mincione et al.[8]	1			1 epithelial neoplasm		
Lack et al.[11]	15	6			9	
Paul et al.[9]	2	2				
Fleming et al.[10]	6	2	2	1 malignant tumor 1 neural neoplasm		
Kapila et al.[12]	3	1		1 adenocarcinoma 1 undifferentiated malignant tumor		
Das et al.[13]	4	3				1
Jashnani et al[14]	1	1				
Majumdar et al[15]	1	1				
Total	52	25	2	12	11	2

nests corresponding to zellballen seen in histology have also been reported.[14] The FNA diagnoses in these cases are depicted in Table 1.[2,7,9,11] Accordingly, the cytologic findings alone, being not specific, are insufficient to provide an initial definitive diagnosis, especially in cases where the clinical findings suggest a different lesion as occurred in our case. Preparation of a cell block from the aspirate on which immunohistochemical studies can also be done is a very useful technique to make a definitive diagnosis. The neoplastic cells will demonstrate strong cytoplasmic positivity with chromogranin indicating neuroendocrine differentiation. The tumor cells also show positivity with synaptophysin, neuron specific enolase and S-100.[9] But in our case the aspirated material was inadequate for cell block processing and hence a definite diagnosis could not be offered. However, in view of the cytologic findings, the possibility of paraganglioma was suggested which was later confirmed in histopathology.

Due to the potential hazards associated with FNA procedure, especially intratumoral hemorrhage and damage to the carotid artery, there is no concordance among authors exposed to FNA of carotid body paragangliomas regarding the need for this investigation in cases where there is a strong clinical suspicion.[1,10] Only one such complication has been reported in the published cases; namely carotid artery thrombosis with resulting cerebral ischemia in Engzell et al.[1] This type of complication could occur in locally aggressive tumors which have invaded the vessel wall, a situation which cannot be assessed at the time of FNA procedure in the absence of precise radiologic studies.[9] Avoiding the carotid vessels can also pose a problem since the anatomic location of the carotid artery and its branches in relation to the tumor mass may vary and in some cases it would be difficult to be determined by mere palpation. The vessels are usually displaced laterally by the tumor, but at times it may widen the carotid bifurcation or displace it medially.[9] Inspite of the limited cytological experience with paragangliomas among pathologists and the rare reported cases of FNA complications, the wider use of FNAC for the diagnosis of such lesions might lead onto the emergence of expected vascular complications.[10, 11] However in clinically obscure cases, FNAC is a simple procedure which would provide a valuable preoperative diagnosis.[14,15]

To conclude, we report a case of carotid body paraganglioma diagnosed on FNAC. The long duration of the swelling, the hemorrhagic nature of the aspirate and the cytomorpholgy of the lesion helped us to suggest this possibility. We would like to highlight the importance of taking a brief clinical history and noting the nature of the aspirate which would be particularly useful in

diagnosing such lesions. We further suggest that preparation of a cell block in cases wherever possible along with the use of immunohistochemical studies would prove useful for the definitive diagnosis of paraganglioma.

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