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Case Report

Extra-dural sporadic sympathetic paraganglioma: A rare care report

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ABSTRACT

Paraganglioma are rare tumor derived from either parasympathetic or sympathetic paraganglia. These tumors have wide spectrum of clinical presentation, and variable anatomic distribution, however their location in the neck and skull base is common but a sporadic location is rare. We are presenting a case of 32 yr female with history of severe systemic hypertension with mid occipital soft tissue swelling. Her radio-imaging study revealed an extra-dural mass lesion with minimal bony erosion. Peroperative course was associated with catastrophic bleeding. On further workup this is found to be a case of sporadic extradural sympathetic paraganglioma, which is not reported in the existing literature.

1. Introduction

Paraganglia are originating from the neural crest that is located throughout the body. Most paragangliomas are relatively slow growing, non-functional and benign; however, some may be malignant or functionally active. Paragangliomas are most commonly associated with hereditary syndrome, often involving Multiple Endocrine Neoplasia type 2A/2B,Von Hippel-Lindau syndrome (VHL) or Neurofibromatosis type 1 (NF1).[1] Spontaneous paragangliomas usually present between the third and fifth decade of life and more commonly in women (71%) than men (29%). Clinical presentation are nonspecific, patient can present with the features of a pheochromocytoma such as hypertension, episodic headaches, diaphoresis, and tachycardia or may be symptomatic due to pressure effect of growing swelling like in our patient. Early suspicion is important because theses lesion can have catastrophic bleeding during surgery and can also leads to various emergencies like hypertensive crisis, seizure disorder and cardiac vascular complications

2. Case Report

A 32-Year old female patient presented with a mid occipital, slowly progressive, mild tender swelling about 4cm x 4cm of eight month duration that was progressive in nature. On general and systemic examination she was having only mild tachycardia HR 104 per minutes and systemic blood pressure was 150 /100, no localizing signs in neurological examination. Rest systemic examinations were within normal limits. She had history of episodic fever off and on. She had history of hypertension which was controlled on medication. We investigated her accordingly and found that CT scan brain suggest a lesion with size of 40x35mm, in mid-occipital area with bony erosion with extradural mass (Figure 1 A, and B). These finding were suggestive of tubercular osteomylitis. Ultrasound guided FNAC done which was nonconclusive and smear was also negative for AFB and Genexpert. She was taken up for surgery with a possible diagnosis of tubercular osteomylitis. Lesion was vascular, 36 x 32 mm and had multiple

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engorged veins over its surface. Patient had copious, diffuse oozing and bleeding during the procedure hence only partial removal could be done and bleeding controlled with difficulty with copious gel foam and pressure application. As the course of disease was unpredictable during the surgery and postoperatively, we planned to evaluate any possibility of metabolically active neuroendocrine tumors. We sent 24hr urinary metanephrines and normetanephrine level for assessment of catecholamine activity of the tumor. The level of normetanephrine was very high 240 nmol/l (normal ranges for normetanephrine in nmol/l < 90 nmol/l) and metanephrines was normal. Tissue was sent for histopathological examination which revealed a circumscribed tumor disposed in diffuse sheets and nested pattern. Individual tumor are polygonal shaped with abundant granular eosinophilic cytoplasm, hyperchromatic nuclei and conspicuous nucleoli, occasional multinucleated cells also seen[Figure 2A-D], the tumor cells showed diffuse positive expression for synaptophysin, chromogranin, loss of expression for S-100 in sustentacular cells and succinate dehydrogenase-B (SDH-B) in tumor cells, low Ki67 proliferation index(1%) and negative expression for pan-cytokeratin [Figure 3A-F], on histopathology and immuno-histochemistry findings a diagnosis of paraganglioma was rendered, since the urinary metanephrines level were high in this patient Grading system for Adrenal Pheochromocytoma and Paraganglioma (GAPP) scoring was also done[2] which included four histological parameter, one immuno-histochemistry and biochemical phenotype, and final diagnosis of sympathetically active paragangliomas, GAPP score = 4(moderately differentiated) was rendered. Further workup done by radio imaging study of abdomen which does not revealed any evidence of other lesion. Patient was comfortable with anti-hypertensive drugs and was referred to endocrinology department for specific therapy of paragangliomas.

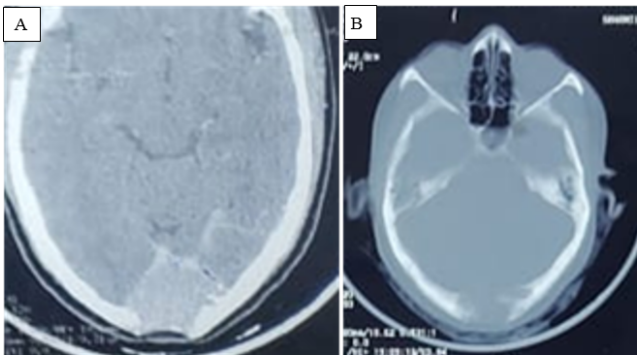

Figure 1: A,B: CT scan brain done to localize and see extant of lesion which showed mid-occipital bony erosion with extradural mass.

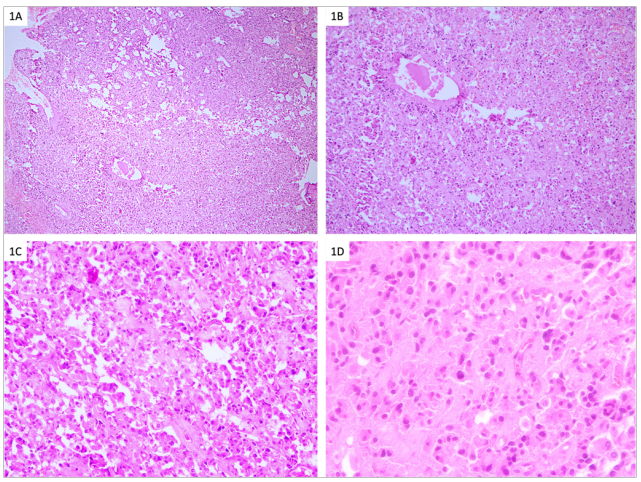

Figure 2: A,B: Photomicrograph showing tumor disposed in diffuse sheets and nested pattern (B,C)Individual tumor are polygonal shaped with abundant granular eosinophilic cytoplasm, hyperchromatic nuclei and conspicuous nucleoli [Hematoxylin and eosin (**A**) 50x (**A**) 100x, (**C**) 200x, (**D**) 400x].

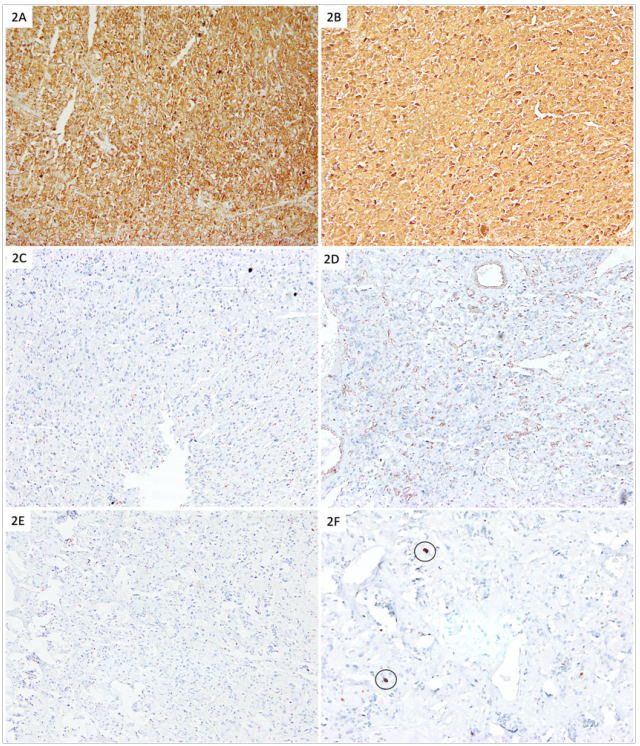

Figure 3: Immunohistochemical findings:**A,B:** Tumor showing diffuse expression for synaptophysin, chromogranin, (**C,D**) loss of expression for S-100 & SDH-B, negative expression for (**E**) Pan-CK & (**F**) low Ki67 proliferation index in tumor [encircled (**E**)]. [Original magnification, (**A-E**) DAB 100x, (**F**)200x]

3. Discussion

Paragangliomas have wide spectrum of anatomical location. They can be found in adrenal and extra-adrenal tissues, and the extra-adrenal paragangliomas can be divided into sympathetic and parasympathetic types[3]. The sympathetic paragangliomas are usually secretory and produce catecholamines while parasympathetic paragangliomas tend to be non-secretory.[4]

In the central nervous system nearly 80–90% paragangliomas occur in the head and neck regions. Sporadic location in rare because these tumor usually occur in association of various endocrinal syndrome. Extradural location in not reported in the literature.

Clinically these tumors present with very nonspecific symptoms that may be due to pressure effect of tumor or active metabolic nature. Metabolically active lesions cause hypertension, palpitation cardiac arrhythmias, sweating and weight loss and pyrexia of unknown origin. Our patient is presented with uncontrolled hypertension due to sympathetic nature of lesion which was confirmed by 24 hours urine normetanephrine level. These tumors are not having any pathognomonic imaging therefore initially misdiagnosed. On CT scans, such lesions appear as homogeneous masses, and rich vascularization.[5] Macroscopically, paragangliomas are a rosy–red to brown color. Microscopically, the principal Type I cells (polygonal, abundant eosinophilic cytoplasm, sometimes presenting granules grouped in nests, so-called 'zellballen') are found surrounded by sustaining Type II cells. Immunohistochemical colorations are essential for a secure diagnosis. The most important neuroendocrine markers are chromogranin and synaptophysin identifies Type I cells and S-100 protein identifies Type II cells[6] in the present case IHC for SDH-B was performed which is poor prognostic indicator and major predictor of metastatic potential in paraganglioma, indicating clinicians a close follow up in such cases. In case of spinal and cauda equina region paragangliomas behave as slow-growing tumors susceptible to potential cure by total excision.[7,8] Elective surgery is a preferred treatment option in most of the cases.

4. Conclusion

Paragangliomas a relatively uncommon tumor that can occur at various anatomical location, their signs and symptoms are nonspecific. Early suspicion is of paramount important for diagnosis and management of these patients. Undiagnosed paragangliomas are associated with unexpected deaths due to operative complications and very high level of catecholamine. There is need to increase awareness regarding these tumors

5. Source of Funding

None.

6. Conflict of Interest

None.

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