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Composite Tumor of the Adrenal Gland-Case Report

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Abstract: Composite-type tumors of the adrenal medulla are rare and account for around 3% of adrenal paraganglioma. Classical features of presentation include hypertension, headache, palpitation and perspiration, while cardiomyopathy and chronic pancreatitis are other reported rarely occurring features of the tumor. Also, many numbers of composite tumors are seen to present with classical signs and symptoms and are solely diagnosed incidentally. The cause behind this lack of catecholamine secretion and thereby lack of clinical classical signs and symptoms of pheochromocytoma especially in composite tumors is largely unknown. One theory says that the ganglioneuroma component might be regulating the catecholamine secretion by pheochromocytoma. It has also been theorized that dopamine production by ganglioneuroma might interfere with the catecholamines secreted by pheochromocytoma. Also, the ganglioneuroma was thought to metabolize the catecholamines produced by pheochromocytoma. We presented a case of a 58-year-old male who was incidentally discovered with a composite adrenal medullary tumor of pheochromocytoma and ganglioneuroma. He did not present with hypertension or any other classical clinical features.

Key words: Composite Tumor · Adrenal Medulla · Pheochromocytoma · Ganglioneuroma

INTRODUCTION

Composite-type tumors of adrenal medulla are rare and might amount to around 3% of adrenal paraganglioma [1-4]. Apparently, they present with clinical features attributed to catecholamine hypersecretion by any of the tumor component [2]. Cardiomyopathy is one of the common presentations of pheochromocytomas [5-7]. Rare presentations of pancreatitis have also been documented [8-12]. Cases with no classical signs and symptoms have also been reported [13]. Diverse nature of presentations might make diagnosis of composite tumors challenging [14]. We recently experienced a case of a composite adrenal medullary tumor of pheochromocytoma and ganglioneuroma that was incidentally discovered and had no signs of catecholamine hypersecretion.

Case Report: A 58-year-old male patient was found to have adrenal mass incidentally and surgical resection of the adrenal gland was done. Greyish brown adrenal tissue en bloc with tumor was measuring 74x67 mm in size. The cut section shows a large cystic lesion, measuring 35x47mm, filled with a pale yellowish, thick, jelly-like content. On further exploration, the adrenal gland shows a brownish appearance of the medulla. Embedded within the periadrenal fat tissue and juxtaposed to it, another well-circumscribed greyish brown lesion, measuring roughly 11x9mm is seen. There is no evidence of capsular invasion.

Microscopically, adrenal gland shows a well-defined capsule and a normal cortex, made up of vacuolated cells relatively exhibiting bland appearance, along with the presence of an unencapsulated paraganglioma lesion involving medulla, measuring 21.13x 3.38 mm. Individual paraganglion cells with abundant brightly eosinophilic cytoplasm, central or eccentrically placed nuclei, arranged in' Zell Ballen 'pattern along with a rich, arborizing, delicate fibrovascular network are noticed (Fig. 1). No pleomorphic features or mitotic activity is seen within this lesion.

Microscopic examination of the well-encapsulated lesion, juxtaposed to the adrenal, showed ganglioneuroma component, measuring 11.03x 9.07 mm displaying abundant ganglion cells, intermixed with nerve fibres in variable proportion (Fig. 2). No apparent atypia or mitotic activity is seen. Cystic lesion showed degenerated pale eosinophilic material and the cyst wall was focally lined by cuboidal cells with stratification in a few areas.

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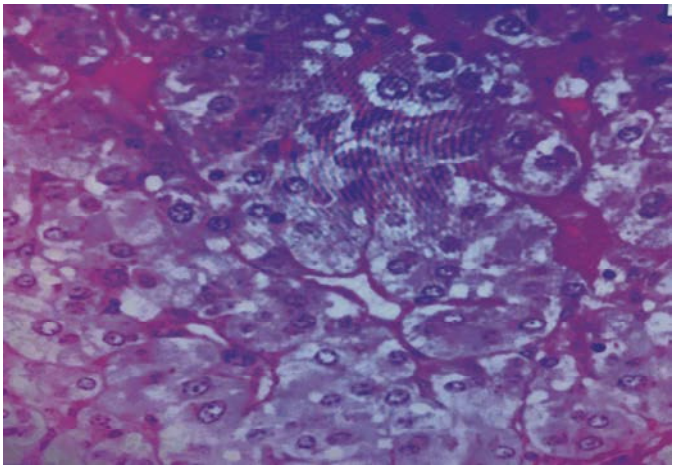

Fig. 1: Shows Pheochromocytoma exhibiting Paraganglion cells in zell-ballen pattern (H&E;20X)

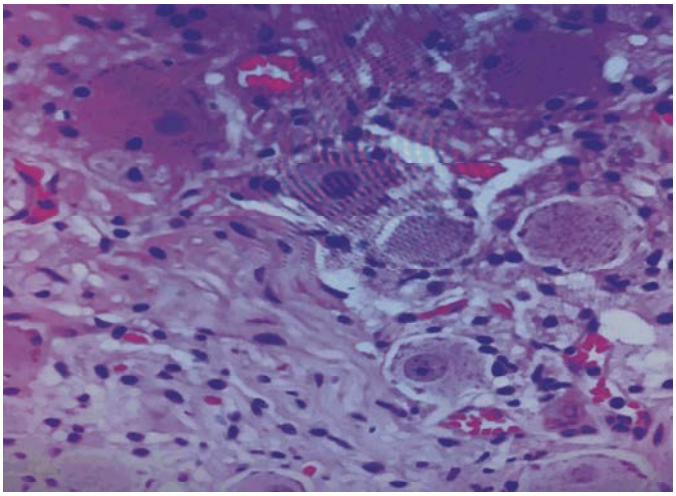

Fig. 2: Shows Ganglioneuroma exhibiting ganglion cells intermixed with nerve fibers (H&E; 20X)

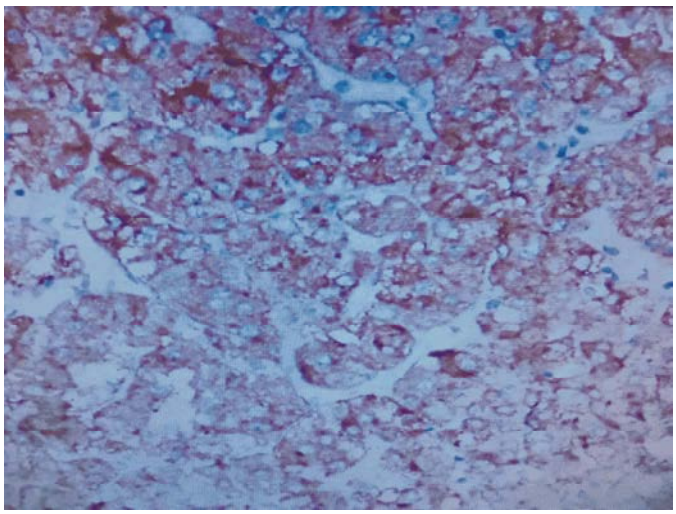

Fig. 3: ShowsSynaptophysin positivity in Pheochromocytoma (Synaptophysin; 20X)

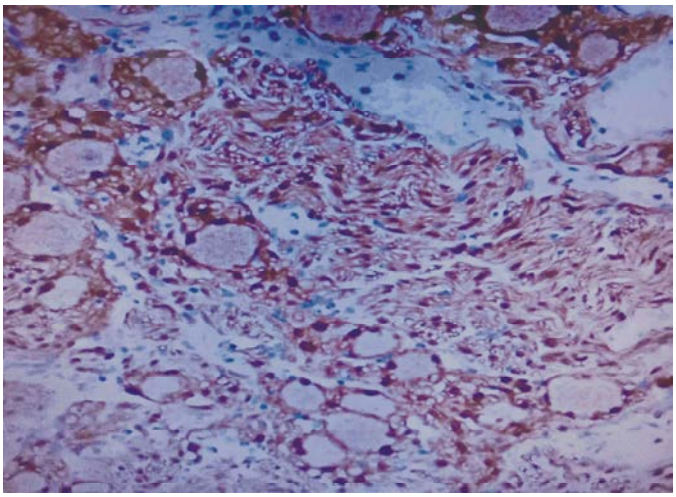

Fig. 4: Shows S100 positivity in Ganglioneuroma (S100; 20X)

Surrounding adipose tissue of the adrenal gland was unremarkable without any evidence of metastatic deposits.

Both paragangliomas of the adrenal gland and juxtaposed ganglioneuroma were studied with an immunohistochemical panel consisting of S100, Synaptophysin, Chromogranin and Ki-67. Strong intense positivity for Synaptophysin was seen within the paraganglioma lesion (Fig. 3) with Ki-67 showing a proliferative index of less than <01% (negligible). Ganglioneuroma showed strong positivity for S-100 but is negative for Chromogranin (Fig. 4).

DISCUSSION

Pheochromocytomas are known to may occur at many different sites and to be in association with a wide variety of other tumors, the occurrence of both pheochromocytoma and ganglioneuroma within a single lesion is very rare. Literature shows very few such cases that have been described. [2-4]. Approximately around 70 cases were known to be reported so far [15]. Pheochromocytomas originate from adrenal chromaffin cells while ganglioneuromas arise from autonomic ganglion cells or their precursors [16], but both chromaffin cells and ganglion cells are derivatives of neural crest cells that migrate to the periphery [17]. This explains the composite appearance of these tumors. Around three-fourths of these tumors present with clinical features of hormonal hypersecretion from either of the tumor components [2, 3]. Headache, palpitation, perspiration are some of the clinical symptoms of secretory pheochromocytomas while sustained or paroxysmal hypertension is the cardinal feature [18].

In our case, the patient had no clinical manifestations of pheochromocytoma attributed to hypersecretion of catecholamines. Literature shows that 72.4% of pheochromocytoma patients present with hypertension, out of which only 47.9% is sustained [14] and the percentage is still lower in composite pheochromocytomas [3]. The cause behind this lack of catecholamine secretion and thereby lack of clinical classical signs and symptoms of pheochromocytoma especially in composite tumors are largely unknown. One theory says that the ganglioneuroma component might be regulating the catecholamine secretion by pheochromocytoma. It has also been theorized that dopamine production by ganglioneuroma might interfere with the catecholamines secreted by pheochromocytoma. Also, the ganglioneuroma was thought to metabolize the catecholamines produced by pheochromocytoma [19].

CONCLUSIONS

Pheochromocytoma can present itself in many different ways without hypertension and other classic clinical features. It is important to identify the rare presenting features of pheochromocytoma in order to prevent an unexpected lethality. We recently experienced a case of a 58-year-old male who was incidentally discovered with a composite adrenal medullary tumor of pheochromocytoma and ganglioneuroma. He did not present with hypertension or any other classical clinical features. Herein, we described the case with a brief review of the literature.

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