This is the case report of 27-year-old woman with presacral teratoma with histopathological features suggestive of paraganglioma (PGLs). This pelvic location of PGLs is fairly uncommon in adults with an incidence of only 2%. On the sonographic examination pelvic PGLs mimics the ovarian masses in consistency and size. The possibility of extra-adrenal pheochromocytoma (PGLs) should be considered in the differential diagnosis.

KEY WORDS: Presacral Teratoma, Pelvic Paraganglioma, Episodic Severe Hypertension, Urinary Retention, Complete Excision.

INTRODUCTION

This is the case report of 27-years-old woman with presacral teratoma with histopathological features suggestive of paraganglioma (PGLs). This pelvic location of PGLs is fairly uncommon in adults with an incidence of only 2%. On the sonographic examination pelvic PGLs mimics the ovarian masses in consistency and size. The possibility of extra-adrenal pheochromocytoma PGLs should be considered in the differential diagnosis. Tumors that originate from adrenal medula are defined as pheochromocytoma whereas tumors located in extra adrenal position are termed PGLs. These PGLs are slow growing, hyper vascular rare neuroendocrine tumors that arise in sympathetic and parasympathetic paraganglion. PGLs are undifferentiated cells of the primitive neural crest, with reported incidence of 1:300 000.

Abdominal PGLs are sympathetic chain derivative tumors found most commonly along aorta in Zuckerkandl body (root of inferior mesenteric artery). Other extra-adrenal sites that have been described include the bladder, pelvis, prostate, ovaries and thorax. Extra-adrenal PGLs can arise anywhere from neck to pelvic floor in locations parallel to sympathetic nervous system. However pelvic presacral location is very rare; only about five cases have been reported so far.

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mass was badly adherent to presacral fascia, retroperitoneum and abutting right ureter but not infiltrating it. The post vesical and post uterine fascia were well defined. Right ureter was separated from mass then tumor was dissected and enucleated from presacral fascia. Uterus, both ovaries and fallopian tubes were normal looking. There was no intra-operative complication. One unit of blood was transfused post-operatively.

Pathologic findings: Gross description: Presacral teratoma consisted of a nodular tissue measuring 6.5 x 5.5 x 5 cm. Microscopic description: revealed an encapsulated tumor with no significant mitotic activity or necrosis (Figure-1). Tumour cells were negative for glycogen (PAS stain). On Immunohistochemical stains Cytokeratin AE1/AE3, Cytokeratin CAM 5.3 and Melan A, were negative however Vimentin, S100 Protein, Chromogranin and Synaptophysin showed the positive reactivity pattern with diagnosis of presacral teratoma: features favouring paraganglioma.

**DISCUSSION**

This case of young female with presacral PGLs is presented because these are rare tumors with reported incidence of 2—8 per million. 11 Sporadic PGLs are usually diagnosed in patients older than 40–50 years, whereas 30% of hereditary forms are diagnosed in younger patients. 4 Although this female was young but no history was suggestive of familial origin of PGLs. PGLs and pheochromocytomas are rare causes of hypertension and only 0.1-1% of all cases of hypertension are due to these tumors. 12 Both PGLs and pheochromocytoma have similar clinical presentation with episodic headache, sweating, tachycardia accompanied with paroxysmal hypertension. Whereas Miss X had sustained hypertension with episodic headache, palpitation and vomiting; this clinical presentation is reported to be more common in children with PGN as reported by Hayes WS et al. 7 One third of patients remain asymptomatic and diagnosis is made by an incidental finding after CT, MRI and ultrasound examination performed for other reasons. 13 Pedroso C. et al 14 and Inazi F. et al 15 reported incidental finding of PGLs during infertility workup in young woman and in another patient during ultrasound abdomen done for suspected nephrolithiasis. In the view of asymptomatic nature of PGLs, high index of clinical suspicion is needed. Scintigraphy (using an analogue of noradrenaline) can help in localization of tumor. 1 Majority of PGLs are non-functional and benign; however, they may secrete excessive catecholamine 16 and may be symptomatic, Miss X also presented with lower abdominal pain, headache, palpitation and severe hypertension. This symptomatic presentation has been reported with hypertensive encephalopathy and hypertrophic obstructive cardiomyopathy with Para aortic invasive paraganglioma 17. Dhiraj KB et all also reported a case of abdominal paraganglioma with hypertensive encephalopathy in 10 year old young boy. 18 Similarly hypertensive emergency during pregnancy 19 and hypertensive crises during thyroidectomy 20 have also been reported in the literature. Total resection is recommended to be curative for PGLs. 21 Complete excision of PGLs in this case was possible, which was evident from postoperative amelioration of clinical signs and symptoms.

**CONCLUSION**

Presacral PGLs are very rare tumors in young women and at times theses mimic ovarian masses on ultrasound. Detailed history and high index of clinical suspicion may prevent mis-assessment/miss-diagnosis. Complete tumor resection may ameliorate the symptoms and be curative.

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**Authors’ Contribution:**

Nusrat Lakho: Conception, designing and manuscript writing
Yasir Masood:Drafting, revising for intellectual contents and literature review.
Shabeen Naz Masood: Final review and edits of the manuscript for publication.

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