Case Study: Extra Adrenal Paraganglioma – Rare Non Gynae Pelvic Mass

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Introduction:

Extra-adrenal paragangliomas (EAPs) are rare nonepithelial neuroendocrine tumours that form near blood vessels and nerves outside of adrenal glands. In contrast intra-adrenal paragangliomas (pheochromocytomas) arise from the adrenal medulla.

About 40% of paragangliomas are inherited. A germline mutation is present in about half of patients presenting with this tumour. 85% of EAPs are in the retroperitoneum. They have high rates of malignancy and require careful histologic and genetic screening for an accurate diagnosis.

Case Report:

A 75 year old female patient was referred by her GP to the Ultrasound Department for a pelvic ultrasound scan. The patient had undergone a PET-CT for a suspicious lung nodule and a central lesion within the pelvis was identified.

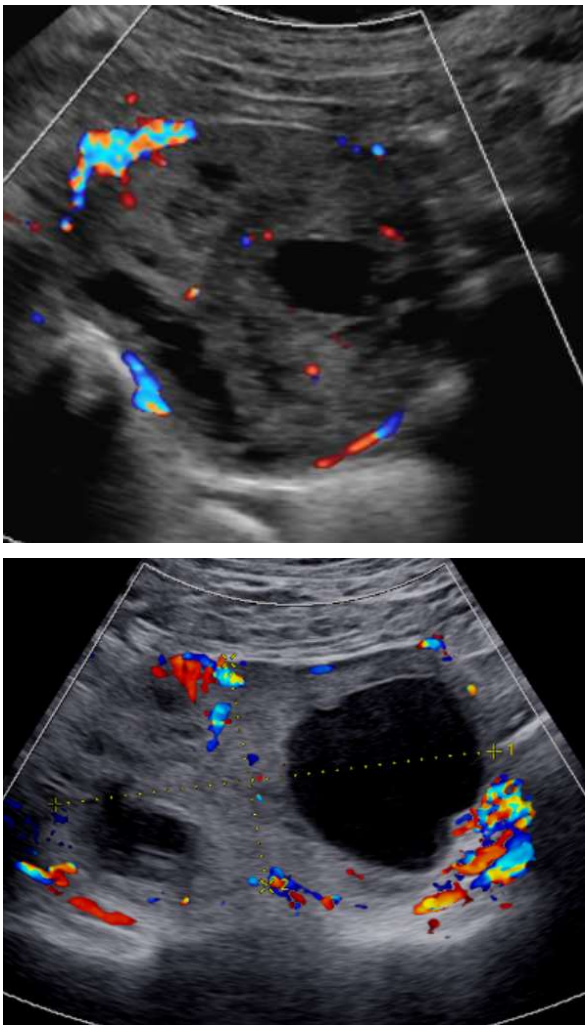

Figure 1: Right Adnexal mass.

A pelvic ultrasound scan was performed. The patient had had a hysterectomy but was unsure if a bilateral oophorectomy had been performed at that time. Grey scale ultrasound images demonstrated (fig 1).

A well-defined solid cystic mass with moderate vascularity measuring 112 x 54 x 73mm was identified within the pelvis. Appearances were suspicious for an ovarian malignancy.

Management

The patient was referred to the Gynaecology Oncology team and her case was discussed at MDT. A detailed review of the patient's medical history verified that a bilateral salpingo-oophorectomy had been performed. A pelvic ultrasound scan two years earlier reported no obvious abnormal findings.

MDT recommended removing the pelvic mass although its origin was uncertain.

A midline laparotomy for excision of the mass was performed. A 12cm fixed retroperitoneal midline mass filling the sacral hollow intimately related to common iliac arteries and ureters bilaterally was identified.

Histopathology analysis reported the following:

1) Infracolic omental biopsy -No evidence of malignancy

2)Retroperitoneal pelvic mass-Extra-adrenal paraganglioma

Discussion:

Extra Adrenal Paragangliomas of the abdomen arise from paraganglia in the retroperitoneum and commonly present in the fourth or fifth decades of life. They are generally regarded as having a poorer outcome than pheochromocytomas with a 20% - 50% incidence of malignancy compared to 10% risk for pheochromocytomas.

Histological features in malignant tumours were not found in this case. Further molecular analysis did not detect a genetic cause

The patient made an excellent recovery and was placed under the care of the endocrinology team and her medical plan is for regular CT abdomen/pelvis surveillance and repeat histology.

References:

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