Answer

Ganglioneuroma

Ganglioneuroma is a rare, benign tumor that is found most commonly in children and young adults and arises from the sympathetic ganglia. It is composed of mature Schwann cells, ganglion cells, and nerve fibers. The posterior mediastinum is the most frequent site of origin of ganglioneuroma. Less frequently it arises in the retroperitoneum and, quite rarely, in the adrenal glands.[1,2]

Characteristically, this tumor does not produce excess catecholamines or steroid hormones,[3] and it usually presents as a clinically silent lesion incidentally detected in imaging studies for unrelated reasons. Surgery is not mandatory if a certain diagnosis of ganglioneuroma is made.

The rarity and the lack of understanding of ganglioneuroma biology often lead to inappropriate diagnosis and/or treatment.[4-6] Unfortunately, there are no specific diagnostic signs or symptoms allowing preoperative discrimination between active ganglioneuroma and pheochromocytoma. In our case, the gradient between plasma catecholamine (normal) and urinary metanephrine levels (elevated) and, furthermore, the atypical appearance on computed tomographic scan (absence of irregular central areas representing necrosis or hemorrhage that, when present in large adrenal masses, are strongly suggestive for pheochromocytoma) most likely should have discouraged the hypothesis of pheochromocytoma. Nevertheless, the large diameter of the lesion was itself a strong indication for surgery because malignancy had to be excluded.

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