Retroperitoneal subduodenal paraganglioma mimicking a GIST : SCARE case

Abstract :

1. Introduction and Importance

Retroperitoneal paragangliomas account for <3% of pheochromocytomas/paragangliomas, are non-secreting in 40-60% of cases, and are often discovered incidentally due to mass effect. A rare case in a 66-year-old woman (outside the peak age range of 40-50 years) mimicking a GIST, without preoperative metanephrines (ESMO/ENSAT IA recommendation omitted). Objective : to highlight the diagnostic pitfall and the value of R0 resection by laparotomy.

2. Case presentation

66-year-old woman, chronic low back pain (history: gastrectomy, cholecystectomy, thyroidectomy). Ultrasound: 47 mm hypoechoic cystic para-umbilical mass. CT scan : 47 mm subduodenal lesion with a hypervascularised necrotic centre suggestive of a GIST. Median laparotomy: R0 resection of a 58 mm polylobular mass (early ligation of vascular pedicles). Histology: benign paraganglioma (chromogranin+/synaptophysin+/S100+/Ki-67<3%/SDHB preserved). Post-operative course : wall infection treated with IV antibiotics, discharged on day 10, followed up at 1 year with no recurrence (negative PET scan).

3. Discussion

Diagnostic pitfall (GIST in 20-25% of incidentalomas >4 cm) without metanephrines (sensitivity >96%, risk of crisis 1-13%). Laparotomy preferable to laparoscopy for vascularised lesions >5 cm (recurrence 3-5% vs 15-20%, OR 4.2).

Limitation: absence of post-operative genetic sequencing (heredity 30-40%).

4. Conclusion

Systematic metanephrine testing for retroperitoneal incidentalomas >4 cm ; laparotomy for vascularised PGLs >5 cm ; genetic sequencing despite benign phenotype (heredity 30-40%). Multidisciplinary management is essential.

***Keywords** : Retroperitoneal paraganglioma, GIST, laparotomy, metanephrines, SCARE, case report*

1. Introduction

Retroperitoneal paragangliomas (RPPG) originate from extra-adrenal sympathetic chromaffin cells. They account for less than 3% of pheochromocytomas/paragangliomas (PPGL) and 6 to 10% of abdominal PPGL (incidence : 1-2/100,000/year) [1,2].

RP-PGLs remain rare (<5% of total PPGLs). They predominantly occur in cephalic sites (85%) and extend from the neck to the pelvic base. Adults show a peak incidence between 40 and 50 years of age. [3–5].

These tumours are functional in 30 to 50% of patients (catecholaminergic secretion: triad of hypertension/sweating/palpitations). Furthermore, 40 to 60% of abdominal forms remain non-secreting. They are often discovered incidentally due to mass effect, with a large volume at diagnosis[5,6].

A hereditary origin affects 30 to 40% of cases (SDHx, VHL, RET genes; MEN2, NF1 syndromes). Clinical latency is long [1, 4,7].

These tumours remain benign in 80 to 90% of cases . Malignancy reaches 10 to 20%, defined by metastases (lymph nodes, bones, lungs, liver). It is associated with an SDHB mutation or Ki-67 >3%, with no reliable histological marker [6,7].

Multidisciplinary assessment precedes curative resection. It explores functionality (plasma/urine metanephrines, sensitivity >96%), extent (CT/PET) and genetics[3,7].

We therefore report an asymptomatic subduodenal PGL-RP, mimicking a GIST, resected in R0 without incident.

2. case presentation

A 66-year-old woman had a history of surgery: gastrectomy for stenotic ulcer, pulmonary tuberculosis treated in 2004, cholecystectomy in 2005, and thyroidectomy for heteromultinodular goitre in 2008.

She consulted the emergency department in November 2024 for chronic low back pain. Clinical examination revealed peri-umbilical tenderness without palpable mass. Blood pressure remained normal, with no cardiovascular or metabolic abnormalities.

Abdominal ultrasound detected a right para-umbilical lesion (47 × 40 × 20 mm), hypoechoic, heterogeneous and cystic, non-vascularised. Abdominal CT confirmed a 47 mm subduodenal retroperitoneal mass, hypodense, heterogeneous with peripheral enhancement and central necrosis (Figure 1). The diagnosis suggested was GIST.

No preoperative biochemical tests (metanephrines) were performed.

Transperitoneal median laparotomy revealed a hypervascularised, multilobulated mass measuring 58 mm in anteposition of the right common iliac vein. R0 resection was performed without Haemodynamic complication, thanks to early ligation of the peritumoral vascular pedicles.

Histology of the specimen confirmed a paraganglioma (chromogranin+, synaptophysin+, S100+, Ki-67 <3%, SDHB preserved). The postoperative period was marked by a wall infection, treated with IV antibiotic therapy. The patient was discharged on postoperative day 10. Follow-up at 1 year showed no recurrence (negative ultrasound and PET scans at 6 and 12 months).

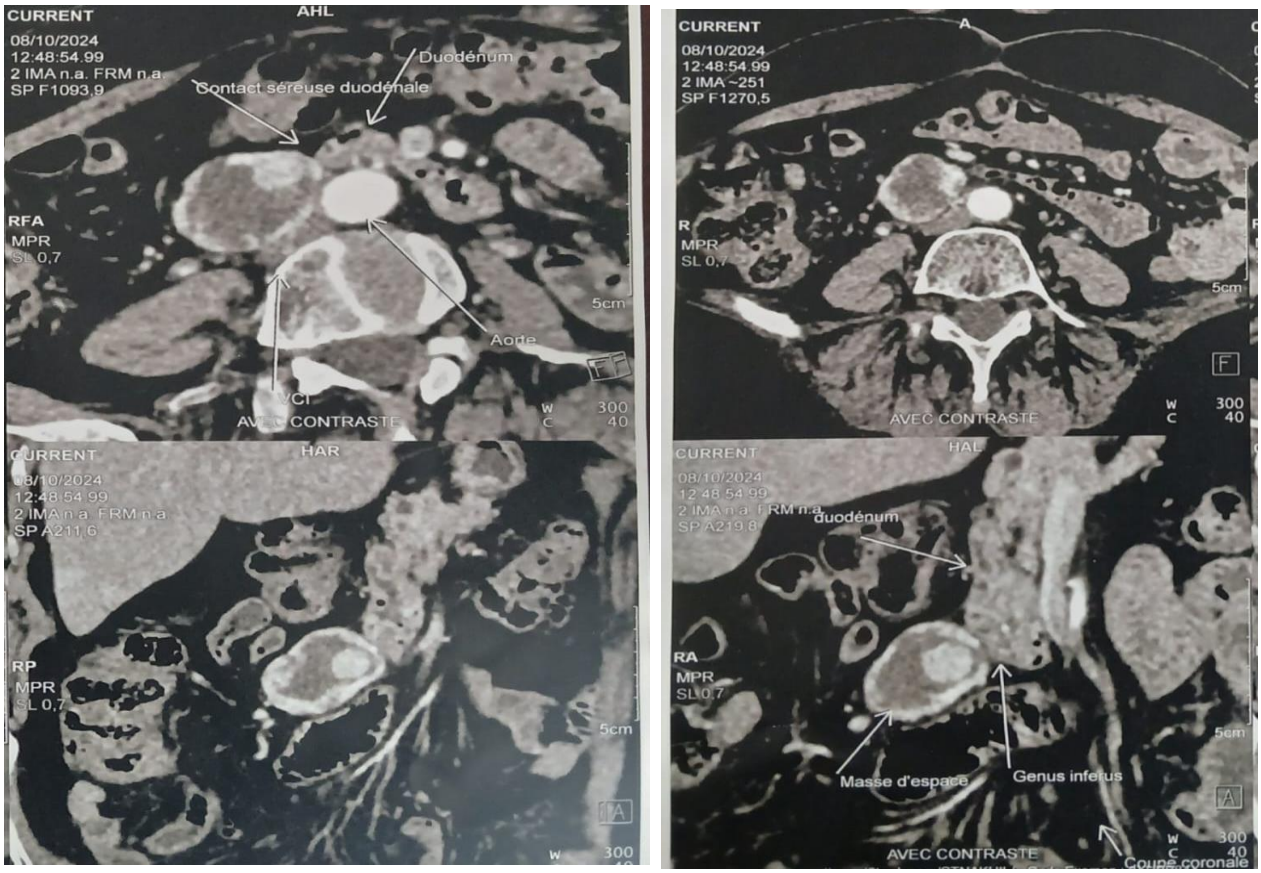

Figure 1 : TDM axiale artérielle : masse tissulaire rehaussante, hétérogène sous-duodénale.

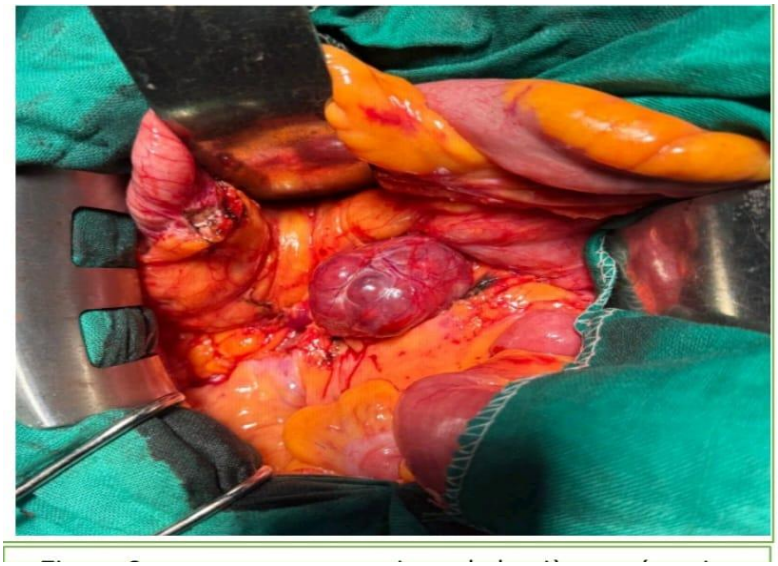

Figure 2 : Aspect macroscopique de la masse en per opératoire

3. Discussion

Retroperitoneal paragangliomas (PGL-RP) originate from sympathetic chromaffin cells. They account for 6 to 10% of abdominal PPGLs (incidence 1-2/100,000/year) [8]. They predominantly occur in the head (85%) [3). Patients typically present between the ages of 40 and 50. The catecholaminergic triad (paroxysmal hypertension, sweating, palpitations) affects 50 to 90% of functional forms. Furthermore, 40 to 60% of abdominal PGLs remain non-secreting[5].

Our observation illustrates a rare variant : isolated chronic low back pain in a 66-year-old woman, outside the peak incidence, accounting for less than 5% of musculoskeletal presentations [1, 2,4].

Ultrasound reveals a 47 mm hypoechoic, cystic, non-vascularised lesion. CT scan shows a hypodense necrotic subduodenal mass with peripheral enhancement, suggestive of a GIST. This differential diagnosis concerns 20 to 25% of retroperitoneal masses larger than 4 cm.

This pitfall affects 15 to 30% of abdominal GISTs due to mass effect. Guidelines recommend

metanephrines (sensitivity >96%) and PET-SSTR for functional characterisation [5, 7,9]. Our non-secreting case confirms this risk of underdiagnosis [5].

We did not measure plasma or urinary metanephrines (grade IA recommendation, ENSAT/ESMO 2023-2024). This omission exposes patients to intraoperative haemodynamic crisis (1-13% without preparation vs <2% with alpha-blockers) [7].

In this case, the non-secreting profile and the absence of the clinical triad explain this decision. However, it contradicts the protocols for retroperitoneal incidentalomas [4]. R0 resection was tolerated without incident, but any incidentaloma larger than 4 cm requires systematic evaluation [10]. The transperitoneal median laparotomy approach allows for careful mobilisation. We ligate the peritumoral pedicles early for curative R0 resection. This strategy is suitable for PGLs larger than 5 cm and vascularised. It reduces local recurrence to 3-5% at 5 years (vs. 15-20% with laparoscopy, OR 4.2) [5,11]. Laparoscopy remains limited to non-vascularised tumours smaller than 5 cm in expert centres (conversion risk 10%) (4].

Immunohistochemistry confirms a benign profile : chromogranin A+, synaptophysin+, S100+, Ki-67 <3%, SDHB preserved. This picture rules out histological malignancy and limits the risk of metastasis to less than 15% (vs. 30-50% for mutated SDHB ; HR 3.5) (12].

PPGLs are hereditary in 30-40% of cases (SDHx 20%, VHL 5-10%, MEN2/NF1). A Post-operative genetic sequencing is required (NGS sensitivity 95%), even without clinical syndrome, for family screening (transmission 15-25%) [1,2]. Its absence is a limitation of our observation.

The prognosis exceeds 95-98% survival at 10 years after R0 resection. Wall infection (5-10% of laparotomies) does not alter this result [13].

4. Conclusion

This atypical subduodenal PGL-RP (66-year-old woman, low back pain mimicking a GIST) highlights three essential surgical points. A systematic biochemical assessment of metanephrines (grade IA ENSAT/ESMO) is imperative for retroperitoneal incidentalomas >4 cm (risk of crisis 1-13%). A median laparotomy is required for vascularised PGLs >5 cm to ensure R0 resection. Finally, postoperative genetic sequencing remains indicated despite a benign phenotype for family screening. Multidisciplinary management optimises diagnosis, complete resection and follow-up of non-secreting PGL-RP.

Consent : Informed consent obtained for publication and figures.

Conflits of interest: The authors declare no conflicts of interest.

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