Multiple endocrine neoplasia type 1 and pancreatic neuroendocrine tumour. Laparoscopic approach



Síndrome de neoplasia endocrina múltiple tipo 1 y tumores neuroendocrinos pancreáticos. Abordaje laparoscópico

Pancreatic neuroendocrine tumours (PNETs) in the context of multiple endocrine neoplasia syndrome (MEN) type 1 are rare, given the low prevalence of this syndrome. Their treatment is still controversial, both in terms of surgical indication and technique¹. Although laparoscopy may offer advantages^{2,3}, the multicentric nature of tumours in MEN 1 syndrome may limit its indications, and few studies have been published in the literature^{4–6}. The aim of this study is to analyse our experience in a tertiary hospital of MEN 1 patients operated on for PNET by laparoscopic approach (1984–2020).

Out of 101 MEN 1 patients, 54% (n = 55) have presented with pancreatic pathology. Sixty-seven per cent (n = 37) underwent surgery, with corporocaudal pancreatectomy (CCP) performed in 53% (n = 36) of cases.

Laparoscopic surgery was performed in 14% (n = 5) of the patients. All had a single tumour in the tail and/or body of the pancreas according to the preoperative morphological study, and underwent splenic-preserving PCC. All patients underwent computed tomography (CT), echoendoscopy and octreoscan, with associated PET-Gallium scintigraphy since 2019. Table 1 shows the characteristics of these patients. The mean follow-up was 86.4 months (24–144). With regard to postoperative complications, patient 1 presented a range of symptoms that was resolved with antibiotic treatment and radiological drainage, and patient 3 presented a pancreatic fistula that was resolved with medical treatment with absolute diet, parenteral nutrition and somatostatin.

Patient 2 also had a 6 cm tumour in the left adrenal gland. The octreoscan image showed a high uptake in the pancreatic tail, with a doubtful uptake in the head-body. No tumour lesion was evident on CT imaging. PCC was performed together with adrenalectomy. At the age of 9 years, a tumour recurrence was seen on the CT scan, in the location where the octreoscan image was captured; the patient was reintervened and an open enucleation was performed.

The benefits of laparoscopic surgery are well known in tumours of the pancreatic body and tail, specifically described in PNETs^{2,3,7}. In MEN 1 patients, it should be evaluated, as these tumours are usually located in the pancreatic tail, although a significant percentage may be multicentric⁸. Few studies have been published on this approach in MEN1^{4–6}. The study by López et al. compares open versus minimally invasive approaches⁵, showing results similar to those described in the meta-analysis by Drymousis et al.² in terms of shorter hospital stay, less blood loss, and no differences in the percentage of pancreatic fistulas⁵.

More than half of the patients had a length of stay of less than one week, less than previously described⁵. In relation to pancreatic fistulas, the percentage described varies from 29% to 62%^{2,5,6}, although no differences are found according to the type of approach, only a higher number of fistulas in cases of enucleation⁹. In our case, the percentage was 20%, although there were no cases with enucleation, and in relation to overall complications, the percentage was similar to that of other studies (40%)².

Another aspect to take into account in our series is that in 100% of the patients it was possible to perform splenic preservation without complications, which seems to be more feasible in minimally invasive surgery⁵.

However, the aspect we consider most important is the possibility of multicentricity and recurrence in these patients⁸. In this regard, the preoperative study is important, as currently, with the introduction of echoendoscopy and Ga PET, sensitivity is close to 100% in tumour detection, and allows us to better select for a laparoscopic approach. In addition, intraoperative ultrasound also allows for better assessment, and is currently a test that we use routinely¹⁰.

Another point to take into account is the functionality or non-functionality of the tumour and the type of secretion. Both non-functioning tumours and insulinomas tend to appear in the pancreatic tail-body, and a laparoscopic

Table 1 – Patients operated on for pancreatic neuroendocrine tumour by laparoscopic approach.										
N	Year	Sex	Age	Site	Type of tumour	Tumour size (cm)	Histology	Stay (days)	F (months)	Rx
1	2018	M	41	Body-tail	NF	3.3	G2	10	24	No
2	2008	M	58	Tail and doubtful uptake in head-body approach	NF	1.8	G1	5	144	Yes
3	2015	M	28	Tail	Insulinoma	2.2	G1	20	60	No
4	2008	W	65	Body-tail	NF	3	G1	6	144	No
5	2015	M	64	Tail	NF	1.8	G1 Multicentre	5	60	No

F: follow-up; G2: grade 1 degree of differentiation: well differentiated; 2: moderately differentiated; Hospital stay; M: man; N: Number of patient; year of surgical intervention; NF: not functioning; Rx: tumour recurrence; W: woman.

approach could be considered, as reflected in our results. However, the indication for more aggressive tumours with a preferential location in the pancreatic head or even duodenum, as in the case of gastrinoma, would be more controversial¹.

In the only study performed in MEN 1 patients comparing recurrence according to the approach, the rate was higher in cases of open surgery, but the follow-up was lower in cases of minimally invasive surgery⁵. On the other hand, it should be noted that the laparoscopic approach in these patients offers the advantage of a lower percentage of postoperative adhesions, which may facilitate surgery in case of reoperation^{2,3,7}.

In conclusion, the laparoscopic approach to pancreatic surgery for non-functioning tumours and insulinomas in patients with MEN 1 is feasible, and can be considered in selected patients with a complete preoperative work-up showing single tumours in the pancreatic corpus coli or multicentric tumours not located in the pancreatic head that allow a laparoscopic PCC to be performed with confidence.

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