Different Management of Pancreatic Head Tumors in Children

Sayed Khedr, MD;¹ Sara Magdy, MD;² Mohamed Saber, MD;¹ Mostafa Gad, MD;¹ Haitham Essmat, MD;¹ Ahmed Arafa, MD¹

¹Department of General Surgery, Faculty of Medicine, Cairo University, Egypt ²Department of General Surgery, Faculty of Medicine, Aswan University, Egypt

Introduction: Less than 0.2% of all juvenile cancer-related fatalities are caused by pancreatic neoplasms, which are uncommon in children.¹

Patients and methods: Review of the literature and a retrospective single-institution research on all child pancreatic tumours during a five-year period. Clinical aspects, presentation, investigation, therapy, and outcome were among the data we provided.

Results: There were five patients found. Abdominal discomfort, mass, and vomiting were the most prevalent symptoms upon presentation. We began oral feeding on the fifth postoperative day; no postoperative complications were discovered; none of our patients had pancreatic or biliary leakage; and all of our cases had free resection margins according to histological analysis.

The neuroendocrine tumour patient and the two SPT cases had pancreaticoduodenectomy.Roux-en-y hepaticojejunostomy and gastrojejunostomy were performed on the other two patients, hemangioendothelioma and rhabdomyosarcoma, who were referred for chemotherapy.

Conclusion: Paediatric pancreatic head tumours are rare, Clinical symptoms are often nonspecific, surgical resection is the optimal treatment in absence of metastatis, Long-term outcome is generally good.

Key words: Pancreatic head mass, hepaticojejunostomy, gastrojejunostomy.

Introduction

Paediatric patients are less likely than adults to develop pancreatic head tumours. Pseudocapillary tumours and neuroendocrine neoplasms are observed in older groups with a fair prognosis, whereas sarcoma had a worse prognosis.^{2,3} Pancreatoblastoma is typical in early childhood.¹

Patients and methods

Five children came to our hospital over 5 years, from march 2016 to march 2021, 2 cases were discovered incidently by abdominal ultrasound, 2 cases presented with obstructive jaundice, CT guided biobsy were done to 4 cases, open biopsy was done in one case.

MRCP was done to cases presenting with obstructive jaundice.

Results

Case Presentation

We present five cases of pancreatic head tumors in children. We found two cases of pancreatic pseudopapillary tumor (SPT), one case of extrahepatic rhabdomyosarcoma, and one case of hemangioendothelioma. The tissues of the fifth case were exceedingly necrotic, and the diagnosis was difficult, but it was probably a neuroendocrine tumor. Pancreaticoduodenectomy was done for the two SPT cases and the neuroendocrine tumor. Rhabdomyosarcoma was referred to chemotherapy. Roux-en-y gastrojejunostomy and hepaticojejunostomy were done for this case and for case of hemangioendothelioma.

Case 1

A fourteen-year-old girl was the initial patient. Over the course of the previous year, the patient had sporadic, nebulous epigastric discomfort that led to hospitalisation. She made no complaints of changed bowel motions or appetite loss. Vital signs were steady. There were no palpable lymph nodes, and she was anicteric. The abdomen was not enlarged, sensitive, or soft. There were no palpable masses or signs of splenomegaly or hepatomegaly. Liver function tests were among the first laboratory results that were essentially normal.

An 8x7.5x7.5 cm pancreatic head tumour was discovered by contrast-enhanced computed tomography (CT) of the abdomen and pelvis **(Fig. 1).** After that, the patient had endoscopic ultrasonography (EUS), which verified the diagnosis. A biopsy was performed on the mass. The patient was subjected to open pancreaticoduodenectomy (Whipple) **(Fig. 2).** There were no problems throughout the postoperative phase. A solid pseudopapillary pancreatic tumour with free surgical margins was identified by histopathological analysis of a well-defined mass of the pancreatic head that measured 8x9x3 cm.



Fig 1: Pancreatic head tumour was discovered by contrast enhanced CT of abdomen and pelvis.



Fig 2: Open pancreaticoduodenectomy (Whipple).



Fig 3: Common Hepatic Duct identification.



Fig 4: Hepaticojejunostomy.



Fig 5: Gastrojejunostomy.

Case 2

A 5-year-old boy was the second patient. The patient was hospitalised after experiencing frequent episodes of vomiting and stomach pain for a month. The patient's vital signs were steady. It was a soft, non-tender abdomen. Hepatosplenomegaly and palpable masses were absent. Alkaline phosphatase was 2224 IU/L (reference range 48.8-445.9 IU/L), total bilirubin was 4.9 mg/dL (Reference range 0.2-1.0 mg/dL), and direct bilirubin was 3.6 mg/dL (Reference range < 0.3 mg/dL), according to the laboratory findings. Roux-en-Y gastrojejunostomy, hepaticojejunostomy, and exploration were performed on the patient (Figs. 3-5). Histopathological analysis revealed a pancreatic primitive sarcomatous tumour that was identified as extrahepatic embryonal rhabdomyosarcoma. Chemotherapy was recommended for the patient. The size of the tumour shrank.

Case 3

A 12-year-old girl complained of frequent episodes of stomach ache. Medical intervention failed to improve the patient's condition. An IV-contrast CT scan of the abdomen and pelvis showed a 4x5 cm tumour near the head of the pancreas that had both solid and cystic components. A biopsy was obtained under the guidance of ultrasonography. pancreatic pseudopapillary neoplasm was Α discovered by histopathological analysis. Open pancreaticoduodenectomy (Whipple) was performed with an uneventful postoperative period. The diagnosis of a pancreatic pseudopapillary tumour was validated by pathological investigation.

Case 4

A male infant, aged 4 months, was brought in with obstructive jaundice. MRCP and abdominal and pelvic MRIs were performed. Alkaline phosphatase was 2500 IU/L (Reference range 81.9-350.3 IU/L), total bilirubin was 8.1 mg/dL (reference

range 0.3-1.0 mg/dL), and direct bilirubin was 5.0 mg/dL (reference range 0.3-1.0 mg/dL).¹⁰ Cholecystostomy and a biopsy of a pancreatic head tumour were performed as part of an abdominal investigation. Roux-en-Y gastrojejunostomy and hepaticojejunostomy were then performed in a second procedure. The postoperative phase went smoothly. After pathological analysis, a vascular spindle cell neoplasm linked to pancreatitis was identified as pancreatic hemangioendothelioma.

Case 5

Contrast and EUS were done. There was an open pancreaticoduodenectomy. The postoperative phase went smoothly. A neuroendocrine tumour was discovered via pathological analysis.

Discussion

Kausch successfully conducted PD for the first time in 1909, and Whipple later spread it in 1935.⁴ Reporting our experience with pancreatic head tumours in children and the indications for pancreaticoduodenectomy surgery is the goal of this retrospective research.

Omar et al,⁵ reviewed the majority of their cases were benign, while malignancy is the majority in our cases.

Grosfeld JL et al,⁶ documented some endocrine pancreatic tumours in neonatal age group, our age cases was 11 years old.

Rebhandl W et al,⁷ reported anaemia and haemoperitoneum presentation in some cases, while in our series, nonspecific symptoms including abdominal pain, vomiting, 2 cases of them, presented with jaundice, two cases were discovered incidently by abdominal ultrasound.

Li D et al,⁸ used stent by ERCP to relieve hyperbilirubinemia in cases of obstructive jaundice, while in our 2 cases of obstructive jaundice, Rouxen-y hepaticojejunostomy and gastrojejunostomy were done.

Hachiyaa M et al,⁹ reported complete regression without the need for surgical intervention in cases of pseudopapillary neoplasm of pancreas, however all our cases needed surgical intervention.

Resende V et al,¹⁰ Zampieri N et al,¹¹ did pancreatectomy with duodenum-preserving approach in cases of benign or low-grade malignant lesions in children with pancreatic head lesions ,while in our resectable cases pancreaticoduodenectomy were done in all 3 cases.

Abete M et al,¹² reported fistula in pancreatic–jejunal anastomosis in their cases with whipple operation, with a 2% to 20% occurrence rate, while in our

cases no fistula was reported

Radical resection of all extrahepatic biliary structures (Including the biliary confluence and the left bile duct) in continuity with all surrounding hilar tissue and the head of the pancreas, as well as partial resection of the portal vein, was reported by Wang KS, et al.,¹³ Giuseppe et al.¹⁴ in cases of rhabdomyosarcoma. The splenomesenteric confluence was used to resect the lower part of the portal vein trunk, direct end-to-end anastomosis was used for vascular reconstruction, and triple bypass Roux-en-y hepaticojejunostomy and gastrojejunostomy were used in our rhabdomyosarcoma patients.

Conclusion

Pancreatic head tumours in children are rare, presentation is typically with non-specific symptoms or as an incidental finding, surgical intervention was advised.

References

- 1. Perez EA, Gutierrez JC, Koniaris LG, et al: Malignant pancreatic tumors: Incidence and outcome in 58 pediatric patients. *J Pediatr Surg.* 2009; 44(1): 197–203.
- Jaksic T, Yaman M, Thorner P, et al: A 20 year review of pediatric pancreatic tumors. *J Pediatr Surg.* 1992; 27: 1315–1317.
- 3. Grosfeld JL, Vane DW, Rescorla FJ, et al: Pancreatic tumors in childhood: Analysis of 13 cases. *J Pediatr Surg.* 1990; 25: 1057–1062.
- 4. Cameron JL, Riall TS, Coleman J, et al: One thousand consecutive pancreaticoduodenectomies. *Ann Surg.* 2006; 244: 10–15.
- Omar N, Nigel JH, Neil JS, Paolo D, Agostino P: Pancreatic tumours in children: Diagnosis, treatment and outcome. *Pediatr Surg Int.* 2015; 31: 831–835.
- Grosfeld JL, Vane DW, Rescorla FJ, McGuire W, West KW: Pancreatic tumors in childhood: Analysis of 13 cases. *J Pediatr Surg.* 1990; 25: 1057–1062.
- Rebhandl W, Felberbauer FX, Puig S, Paya K, Hochschorner S, Barlan M, Horcher E: Solidpseudopapillary tumor of thepancreas (Frantz tumor) in children: Report of four cases and review of the literature. *J Surg Oncol.* 2001; 76: 289–296.
- 8. Li D, Xie K, Wolff R, Abbruzzese JL: Pancreatic cancer. *Lancet*. 2004; 363: 1049–1057.
- 9. Hachiyaa M, Hachiyaa Y, Mitsuia K, TsukimotoI, Watanabe K,Fujisawa T: Solid, cystic and

vanishing tumors of the pancreas. *J Clin Imag.* 2003; 27: 106–108.

- 10. Resende V, Azevedo PH, Lima Ldo P, Portela AR, Sanches MD, Pedrosa MS: Duodenumpreserving pancreatic headresection in solid pseudopapillary neoplasm-report of a case. *Int J Surg Case Rep.* 2014; 5: 567–570.
- 11. Zampieri N, Schiavo N, Capelli P, Scarpa A, Bassi C, Camoglio FS: Pseudopapillary tumor in pediatric age: Clinical and surgical management. *Pediatr Surg Int.* 2011; 27: 1271–1275.
- 12. Abete M, Ronchetti V, Casano A, et al: Pancreatic

fistula after pancreaticoduodenectomy: Risk factors and treatment. *Minerva Chir.* 2005; 60: 99–110.

- 13. Wang KS, Albanese C, Dada F, et al: Papillary cystic neoplasm of the pancreas: A report of three pediatric cases and literature review. *J Pediatr Surg.* 1998; 33(6): 842–845.
- 14. Giuseppe d'Ambrosio, Laura del Prete a, Chiara Grimaldi a, Arianna Bertocchini, Cristina Lo Zupone, Lidia Monti b, Jean de Ville de Goyet: Pancreaticoduodenectomy for malignancies in children. *Journal of Pediatric Surgery*. 2014; 49: 534–538.