Pancreatic involvement in Erdheim-Chester disease: a case report and review of the literature

Jia-wen Dai², Tian-hua He², Ming-hui Duan², Yue Li³ and Xin-xin Cao¹,2*

Abstract

**Background:** Erdheim-Chester disease (ECD) is a rare form of non-Langerhans cell histiocytosis characterized by infiltration of lipid-laden foamy macrophages within different tissues. Clinical manifestations of ECD are highly heterogeneous. Bone lesions are found in 80%-95% of patients, while extraosseous lesions usually involve the cardiovascular system, retroperitoneum, central nervous system (CNS), and skin. Pancreatic involvement in ECD has barely been reported.

**Case presentation:** A 29-year-old female initially presented with menoxenia, diabetes insipidus and diabetes mellitus. 18F-fluorodeoxyglucose positron emission tomography-computed tomography (18F-FDG-PET/CT) revealed hypermetabolic foci in the bilateral frontal lobe, saddle area, and pancreas. A 99mTc-MDP bone scintigraphy scan revealed symmetrical increased uptake in distal femoral and proximal tibial metaphysis, which was confirmed to be osteosclerosis by high-resolution peripheral quantitative computed tomography. The patient underwent incomplete resection of the sellar mass. Histological examination of biopsies showed histiocytic aggregates, which were positive for S100 and negative for CD1a and CD207 on immunohistochemistry. Enhanced abdominal CT scan showed hypointense nodules within the body and tail of the pancreas. Endoscopic ultrasonography guided fine-needle aspiration (EUS-FNA) found no evidence of malignancy. She was diagnosed with ECD and treated with high-dose IFN-α. Repeated examinations at three-and eight-months post treatment revealed markedly reduction of both intracranial and pancreatic lesions.

**Conclusions:** ECD is a rare histiocytic neoplasm that can involve almost every organ, whereas pancreatic involvement has barely been reported to date. Here, we present the rare case of pancreatic lesions in ECD that responded well to interferon-α. We further reviewed reports of pancreatic involvement in histiocytic disorders and concluded the characteristics of such lesions to help diagnosis and treatment, in which these lesions mimicked pancreatic adenocarcinoma and caused unnecessary invasive surgeries.

**Keywords:** Pancreas, Histiocytosis, Erdheim-Chester disease, Treatment, Interferon, Case report

---

**Background**

Erdheim-Chester disease (ECD) is an inflammatory myeloproliferative neoplasm characterized by infiltration of tissues by foamy CD68⁺CD1a⁻ histiocytes [1]. Theoretically, ECD can affect every tissue and organ, while so far pancreatic involvement has been reported only in one case. The main sites of involvement in ECD patients include bone (95%), lung (91%) [2], cardiovascular region...
(50%), retroperitoneum (40–50%), central nervous system (40%), and skin (25%) [3]. Iconic radiographic signs of ECD include the ‘hairy kidney’, sheath around the aorta, long-bone sclerosis, and right atrial pseudo tumors. Clinical manifestations can be of great heterogeneity. Any of the clinical signs, such as bone pain, diabetes insipidus, xanthelasma, exophthalmos, ataxia, or sinusitis, may herald the disease [4]. The mean time from symptom onset to diagnosis was 2.7 years [5]. Mutations activating the MAPK pathway are found in more than 80% of patients with ECD, mainly the \( \text{BRAF}^{V600E} \) mutation in 57% to 70% of cases, followed by MAP2K1 in close to 30% [1, 6–9]. Untreated multisystemic ECD can be severe and fatal. Patients with life-threatening cardiac or neurologic involvement with or without \( \text{BRAF-V600E} \)-mutation should receive MEK inhibitors. For \( \text{BRAF-wild-type} \) patients without end-organ dysfunction, IFN-\( \alpha \) is still the first line therapy, especially in developing countries. A retrospective cohort study reported a response rate of 80%, and 3-year progression-free survival and overall survival of 64.1 and 84.5%, respectively [10]. \( \text{BRAF} \) and MEK inhibitors have shown robust efficacy in \( \text{BRAF}^{V600E} \) patients, yet most patients relapsed after \( \text{BRAF inhibitor interruption} \) [11]. ECD involving the pancreas has barely been reported. Our case highlights a rare location, the pancreas, for a rare disorder, Erdheim-Chester disease. We also reviewed reported cases of pancreatic involvement in relatively common histiocytic disorders for better diagnosis and management, including Langerhans cell histiocytosis (LCH), Juvenile xanthogranuloma (JXG), and Rosai-Dorfman disease (RDD).

**Case presentation**

A 29-year-old female presented to our hospital with a complaint of menoxenia for 5 years and polyuria, polydipsia, hyperglycemia and lethargy for 1 year, with no previous medical, family, and psycho-social history. She was diagnosed with menoxenia in 2013 and treated with hormone replacement therapy. In 2017, when she gradually developed symptoms of diabetes insipidus and lethargy, a brain MRI was arranged which showed a mass in sellar area. Incomplete resection was performed, and histological examination of the mass showed histiocytic aggregates, which were CD1a-negative, Langerin-negative, and S100-positive on immunohistochemistry. (Fig. 1a, b. Microscope: OLYMPUS BX53; acquisition software: pylon Viewer; measured resolution: 1390*1038px; scale bar: 50 \( \mu \)M). DNA extracted from the patient’s biopsy sample was obtained and subjected to NGS of 183 genes, including \( \text{BRAF}, \text{MAP2K1}, \text{PIK3CA}, \text{NRAS}, \text{KRAS}, \text{ARAF}, \text{ALK} \) [9], yet no \( \text{BRAF}^{V600E} \) and other meaningful mutations downstream the MAPK or in related pathways was found.

2 years later, the patient was admitted to our hospital due to progression of the intracranial mass. We performed further examinations to confirm the diagnosis. On physical examination, no remarkable abnormality was found. Blood test and tumor markers were normal. Liver enzymes were abnormal with a mild to moderate elevation of alkaline phosphatase (178 U/L) and \( \gamma \)-Glutamyltransferase (72 IU/L). C-reactive protein, erythrocyte sedimentation rate, and Tumor necrosis factor-\( \alpha \) elevated slightly. Enhanced MRI of the brain showed multiple lesions affecting sella, suprasellar area, pons, and part of hypothalamus (Fig. 1c). The patient’s 99mTc-MDP bone scintigraphy scan revealed symmetrical increased uptake in the frontal bone and distal femoral and proximal tibial metaphysis (Fig. 1d).

Further investigation with high-resolution peripheral quantitative computed tomography (HR-pQCT) confirmed long-bone osteosclerosis by revealing increased trabecular volumetric bone mineral density and localized structural alteration of trabecular network in tibia (Fig. 1e) [12]. FDG-PET/CT revealed hypermetabolic foci in the bilateral frontal lobe, nasal septum, sella, gallbladder, and the body and tail of the pancreas (Fig. 1f). Further examination on the pancreas with enhanced CT scan showed nodules of hypointense lesions within the body and tail of the slightly enlarged pancreas. During the arterial phase and portal phase, such lesions showed reduced enhancement (Fig. 1g).

Based on typical meta-diaphyseal osteosclerosis and pathological findings of histiocytes aggregates, the patient was diagnosed with ECD, involving the brain, bones and the pancreas. She was treated with IFN-\( \alpha \) at 900 million international units, three times a week. Hormone replacement therapy included euthyrox and minirin. Metformin was also applied to control blood glucose. She tolerated the treatment well with no unanticipated events. Repeated MRI of the brain at three- and eight-months post treatment showed alleviation of all intracranial lesions (Fig. 2b, c). Repeated abdominal CT scans revealed markedly reduction of size of the pancreatic lesions, and their enhancement features were closer to normal pancreatic tissue (Fig. 2e, f). The patient still relied on hormone replacement therapy but her lethargy largely resolved, and her blood glucose level was easier to control.
Discussion and conclusion

In this case, though EUS-FNA of pancreas found no evidence of infiltration of histiocytes, those nodular, obscure circumstanced, hypermetabolic lesions, with rather a rapid response to IFN treatment, were suggested as ECD involvements. We should consider pancreatic tumor, chronic pancreatitis, and autoimmune pancreatitis in those space-occupying lesions, of which we are most concerned about pancreatic tumor. However, the lesions were not accompanied by indirect signs of malignancy such as ductal dilation and vascular invasion, tumor markers are normal, and no tumor cells were found by pathological biopsy, thus we excluded this diagnosis.

The histiocytoses are rare disorders characterized by the accumulation of macrophage, dendritic cell, or monocyte-derived cells in various tissues and organs. Histiocytic disorders were traditionally divided into Langerhans cell histiocytosis (LCH) and non-Langerhans cell histiocytosis, among which Erdheim-Chester disease (ECD), Juvenile xanthogranuloma (JXG), and Rosai-Dorfman disease (RDD) were the most common types. Since pancreatic involvement is rare in histiocytoses, we know little about the characteristics of such lesions. Thus, we searched case reports of histiocytoses involving pancreas in the English literature in the PubMed database. Thus far, only one pancreatic ECD has been reported.

Fig. 1 Imaging and pathological data at the time of diagnosis. a Histological examination of the sellar mass showed histiocytic aggregates (x 200, scale bar: 50 µM), which were CD1a-negative, Langerin-negative, and b S100-positive on immunohistochemistry (x 200, scale bar: 50 µM). c Enhanced brain MRI showed multiple lesions affecting sellar, suprasellar area, pons, and part of hypothalamus. d ⁹⁹mTc-MDP bone scintigraphy scan showed symmetrical increased uptake in the frontal bone and distal femoral and proximal tibial metaphysis. e HR-pQCT confirmed osteosclerosis by revealing increased trabecular volumetric bone mineral density and localized structural alteration of trabeculae network in tibia. f PET/CT revealed hypermetabolic foci in the bilateral frontal lobe, saddle area, and pancreas. g Enhanced abdominal CT scan showed nodules of hypointense lesions within the body and tail of the slightly enlarged pancreas. h EUS-FNA of pancreas found no evidence of malignancy but only normal pancreatic ductal cells (x 400, scale bar: 25 µM).
while 5 cases of LCH (Table 1), 19 cases of JXG (Table 2), and 11 cases of RDD (Table 3) have been reported. In the following tables, we summarized the key information of these cases.

Pancreatic involvement in ECD was reported in a 57y woman with pancreatic induration, which was confirmed of ECD involvement by biopsy. The patient died of acute respiratory failure of unknown cause 5 months later [13]. All of 5 cases of LCH were high risk, with involvement in the liver, spleen, or bone marrow. All patients received chemotherapy, but the condition was resolved in only 2 patients. The third patient showed an exact size reduction of the pancreatic lesion, similar to what we reported in our case. It is reasonable to believe the pancreas is involved more often in high-risk LCH. The 19th case of JXG was a baby with a lesion in the head of the pancreas and largely elevated cancer antigen 19-9 (1954 U/mL). She underwent Whipple surgery

![Imaging Changes of the Brain and Pancreas During Treatment](image)

**Fig. 2** Imaging changes of the brain and pancreas during treatment. Enhanced brain MRI scans at **a** pre-treatment, **b** three months, and **c** eight months post treatment. Enhanced abdominal CT scans of pancreas at **d** pre-treatment, **e** three months, and **f** eight months post treatment.

Table 1 Summary of 5 cases with ECD and LCH involving the Pancreas

| No. | References     | Sex/age | Symptoms                  | Site          | Treatment | Outcome | Other organs involved             |
|-----|----------------|---------|---------------------------|---------------|-----------|---------|-----------------------------------|
| 1   | Poehling et al. [13] | F/57y   | Cramping                  | Autopsy       | Prednisone| Death   | Bone, kidney                     |
| 2   | Hara et al. [16]   | M/10y   | Fever, jaundice           | Diffuse swelling | Chemo (EP) | Death   | Lung, liver, spleen, BM, kidney   |
| 3   | Yu et al. [17]     | M/8mo   | Belly pain, distension, diarrhea | Autopsy       | Chemo (VP, C) | Death   | Skin, liver, spleen, BM, lung, GI |
| 4   | Muwakkit et al. [18] | M/4w    | Frequent stools           | Body (cyst)   | Chemo (VP) | Resolution | Skin, lung, spleen               |
| 5   | Goyal et al. [19]  | M/18mo  | Loose stools              | Autopsy       | Chemo (VP) | Death   | LN, liver, kidney                |
| 6   | Hou et al. [20]    | M/44y   | /                         | Diffuse swelling | Chemo (CAVP) | Resolution | Lung, liver, LN, bone            |

BM bone marrow, GI gastrointestinal tract, LN lymph node, Chemo chemotherapy, E etoposide, V vinblastine, P prednisone/prednisolone, C cyclosporin A, A adriamycin
as a diagnostic and therapeutic method and resolved well, with normalization of CA 19-9 within 1 month. Such lesions, especially those with elevated tumor markers, are difficult to differentiate with malignancies. From these cases, we can conclude that the symptoms of the over 30 cases mentioned are quite atypical, ranging from obstructive jaundice to no discomfort. The pancreas can be affected in different forms, with solid

| No. | References | Sex/age | Symptoms | Site | Treatment                  | Outcome | Other organs involved |
|-----|------------|---------|----------|------|---------------------------|---------|-----------------------|
| 1   | Dehner [21]| M/2mo   | Jaundice | Head | Unknown                   | Resolution | Lung                  |
| 2   | Heintz et al. [22]| F/5mo | Jaundice | Head | Whipple                   | Resolution | Liver                 |
| 3   | Prasil et al. [23]| NA/9mo | Jaundice | Head | Mass excision             | Resolution | –                     |
| 4   | Ueno et al. [24]| M/42y | Belly pain | Body (cyst) | Distal pancreatectomy     | Resolution | –                     |
| 5   | Iyer et al. [25]| M/50y | Jaundice | Head | Whipple                   | Unknown   | Unknown               |
| 6   | Iyer et al. [25]| M/36y | Pancreatitis | Tail | Mass excision             | Unknown   | Unknown               |
| 7   | Kamitani et al. [26]| M/82y | Belly pain | Body (cyst) | Whipple                   | Unknown   | Stomach               |
| 8   | Kang 2007 | F/22y | Belly pain | Head | PPPD                      | Unknown   | Unknown               |
| 9   | Okabayashi et al. [27]| M/60y | Belly pain | Tail | Distal pancreatectomy     | Unknown   | Unknown               |
| 10  | Okabayashi et al. 2007 | M/60y | Belly pain | Tail | Distal pancreatectomy     | Unknown   | Unknown               |
| 11  | Shima et al. [28]| M/66y | Belly pain | Body | Distal pancreatectomy     | Unknown   | –                     |
| 12  | Iso et al. [29]| M/82y | Weight loss | Head and tail | Distal pancreatectomy     | Resolution | Spleen               |
| 13  | Ikeura et al. [30]| M/73y | – | Body (cyst) | PPPD                      | Unknown   | –                     |
| 14  | Uguz et al. [31]| M/30y | Belly pain | Head | PPPD                      | Unknown   | Unknown               |
| 15  | Uguz et al. [31]| M/34y | Belly pain | Head | PPPD                      | Unknown   | Unknown               |
| 16  | Kim et al. [32]| F/72y | Weight loss | Body (cyst) | PPPD                      | Resolution | –                     |
| 17  | Kim et al. [33]| F/70y | Belly pain, dyspepsia | Uncinate | PPPD                      | Resolution | –                     |
| 18  | Antary et al. [34]| M/60y | Belly pain, vomit | Uncinate | PPPD                      | Resolution | –                     |
| 19  | Antary et al. [35]| F/13mo | Jaundice | Head and uncinate | Whipple       | Resolution | –                     |

**PPP D** pylorus preserving pancreatectoduodenectomy

| No. | References | Sex/age | Symptoms | Site | Treatment                  | Outcome | Other organs involved |
|-----|------------|---------|----------|------|---------------------------|---------|-----------------------|
| 1   | Esquivel et al. [36]| F/48y | Belly pain | Body and tail | Distal pancreatectomy     | Unknown   | Spleen               |
| 2   | Zivin et al. [37]| F/63y | Jaundice | Body | Whipple                   | Resolution | Lung                 |
| 3   | Podberezin et al. [38]| F/35y | Belly pain | Tail | Mass excision             | Progression (steroids, chemo, imatinib, excision) | Spine, perinephric, perisplenic |
| 4   | Romero et al. [39]| F/74y | Belly pain | Head | PPPD                      | Unknown   | –                     |
| 5   | Shaikh et al. [40]| F/59y | Belly pain | Body and tail | Whipple, steroids         | Progression (imatinib) | Liver            |
| 6   | Mantilla et al. [41]| F/54y | Belly pain, weight loss | Tail | Distal pancreatectomy     | Resolution | –                     |
| 7   | Karajikar et al. [42]| F/65y | Belly pain | Head, body, and tail | Consider clofarabine | Unknown   | Presacral soft tissue, skin |
| 8   | Smith et al. [43]| F/75y | Weight loss | Body | Steroids                  | Resolution | –                     |
| 9   | Brown et al. [44]| F/65y | Granulomatous uveitis, skin rash | Tail | Distal pancreatectomy     | Resolution | Skin                 |
| 10  | Liu et al. [45]| F/71y | Fullness | Tail | Distal pancreatectomy     | Resolution | –                     |
| 11  | Emily et al. [46]| F/40y | Belly pain | Tail | Distal pancreatectomy     | Resolution | Colon                |
or cystic masses in the head/body/tail or diffuse swelling of the whole pancreas. It can be involved in the disease alone or with any possible organ. Due to the similarity in clinical presentation and imaging with pancreatic malignancies, these lesions mostly lead to distal pancreatectomy or even Whipple surgery, with only one patient among all 30 cases of JXG and RDD receiving medical treatment.

However, considering the spontaneous remission trend of JXG and RDD and the good response of these two diseases as well as LCH and ECD to chemotherapy or targeted BRAF inhibitors, we believe that surgery is sometimes overprescribed to a certain extent. Therefore, histiocytoses may be considered as a differential diagnosis for patients presenting with a pancreatic mass.

Recently, two recent publications have explained the cause of the hyperinflammatory state in ECD and other histiocytic diseases. Molteni, R. and his colleagues found that BRAFV600E in macrophages induce hallmark immunometabolic features of trained immunity, causing activation of the AKT/mTOR signaling axis, increased glycolysis, epigenetic changes on promoters of genes encoding cytokines, and enhanced cytokine production leading to hyper-inflammatory responses [14]. Biavasco, R. and his colleagues discovered that the activation of BRAFV600E impairs HSPC function, features myeloid restricted hematopoiesis, and leads to a widespread inflammatory condition [15]. These findings reveal the cause of high inflammatory condition in ECD patient, explain the rationale for pancreatic involvement and the robust response to IFN in our case.

In conclusion, we report the second case of pancreatic ECD with a good response to interferon-α therapy, with a literature review of pancreatic involvement in other histiocytoses, including LCH, JXG, and RDD. These lesions often simulate pancreatic malignancies, causing unnecessary invasive surgery in some cases. Thus we recommend histiocytoses as a differential diagnosis in pancreatic lesions.

**Abbreviations**

ECD: Erdheim-Chester disease; CNS: Central nervous system; 18F-FDG-PET/CT: 18F-fluorodeoxyglucose positron emission tomography-computed tomography; HR-pQCT: High-resolution peripheral quantitative computed tomography; L: Lymph node; RDD: Rosai-Dorfman disease; NGS: Next Generation Sequencing; BM: Bone marrow; GI: Gastrointestinal tract; THH: and YL analyzed relevant information, JWD performed literature review and wrote the manuscript; MHD, YL, and XXC clinically managed the patient. All Authors read and approved the final manuscript.

**Funding**

Institutional research funding was provided by the Innovation Training Program for College Students of Peking Union Medical College [XE100000100090]. The funding body played no role in the design of the study and collection, analysis, and interpretation of data and in writing the manuscript.

**Availability of data and materials**

The data used and analyzed during the current study are included in this article.

**Declarations**

**Ethics approval and consent to participate**

Not applicable.

**Consent for publication**

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the editor of this journal.

**Competing interests**

The authors declare that they have no competing interests.

**Author details**

1 Department of Hematology, State Key Laboratory of Complex, Severe and Rare Diseases, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China. 2 Department of Hematology, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China. 3 Department of Gastroenterology, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China.

**Received:** 3 February 2022   **Accepted:** 10 June 2022

**Published online:** 21 June 2022

**References**

1. Emile JF, Abla O, Fraitage S, Horne A, Haroche J, Donadieu J, Requena-Caballero L, Jordan MB, Abdel-Wahab O, Allen CE, et al. Revised classification of histiocytoses and neoplasms of the macrophage-dendritic cell lineages. Blood. 2016;127(22):2672–81.
2. Wang JN, Wang FD, Sun J, Liang ZY, Li J, Zhou DR, Tian X, Cao XX. Pulmonary manifestations of Erdheim-Chester disease: clinical characteristics, outcomes and comparison with Langerhans cell histiocytosis. Br J Haematol. 2021;194(6):1024–33.
3. Goyal G, Young JR, Koster MJ, Tobin WQ, Vassallo R, Ryu JH, Davidge-Pitts CJ, Hurtado MD, Ravindran A, Sartori Valinotti JC, et al. The Mayo Clinic Histiocytosis Working Group consensus statement for the diagnosis and evaluation of adult patients with histiocytic neoplasms: Erdheim-Chester disease, Langerhans cell histiocytosis, and Rosai-Dorfman disease. Mayo Clin Proc. 2019;94(10):2054–71.
4. Diamond EL, Dagna L, Hyman DM, Cavalli G, Janku F, Estrada-Veras J, Ferrarini M, Abdel-Wahab O, Heaney ML, Scheel PJ, et al. Consensus guidelines for the diagnosis and clinical management of Erdheim-Chester disease. Blood. 2014;124(4):483–92.
5. Haroche J, Cohen-Aubart F, Amoura Z. Erdheim-Chester disease. Blood. 2020;135(16):1311–8.
6. Cao XX, Sun J, Li J, Zhong DR, Niu N, Duan MH, Liang ZY, Zhou DB. Evaluation of clinicopathologic characteristics and the BRAF V600E mutation
in Erdheim-Chester disease among Chinese adults. Ann Hematol. 2016;95(5):745–50.

7. Badalani-Very G, Vergilio JA, Degar BA, MacConaill LE, Brandner B, Calic-Dai R, Poehling GG, Adair DM, Haupt HA. Erdheim-Chester disease. A case report. Clin Orthop Relat Res. 1984;185:241–4.

8. Cao XX, Niu N, Sun J, Cai H, Wang Z, Zhao AL, Duan MH, Zhou DB, Li M, Liu F, Shen J, Yin J, Wu S, Lu F, Jia W. Adult multisystem Langerhans cell histiocytosis involving parathyroid glands and pancreas. Chin Med J (Engl). 2014;127(8):1597.

9. Cen J, Zhao AL, Duan MH, Cao XX, Niu N, Sun J, Cai H, Wang Z, Zhao AL, Duan MH, Zhou DB, Li M, Liu F, Shen J, Yin J, Wu S, Lu F, Jia W. Adult multisystem Langerhans cell histiocytosis involving parathyroid glands and pancreas. Chin Med J (Engl). 2014;127(8):1597.

10. Chib N, Ceylan B, Anand S, Lyonar R, Parikh S, Cai H, Jiang Y, et al. Bone mineral density and bone microarchitecture in a cohort of patients with Erdheim-Chester disease. Orphanet J Rare Dis. 2020;15(1):236.

11. Dahanau J, Kim E, Cohen-Aubart F, et al. Diverse and targetable kinase alterations in adult histiocytosis. Leukemia. 2022;36(2):573–6.

12. Dehner LP. Juvenile xanthogranulomas in the first two decades of life: a clinicopathologic study of 174 cases with cutaneous and extracutaneous manifestations. Am J Clin Pathol. 2003;120(5):579–93.

13. Dong H, Shi L, Shi L, Liu Y, Zou X, et al. Xanthogranulomatous pancreatitis associated with intraductal papillary mucinous carcinoma. J Gastrointest Cancer. 2012;43(4):626–9.

14. Gupta K, Amin M, Heinz J, Voo K. Xanthogranulomatous pancreatitis: third reported occurrence. J Gastrointest Cancer. 2012;43(4):626–9.

15. Harris SR, Fullagar KL, Caridi J, Lewis IR, Cheung H, Clamp CP. Xanthogranulomatous pancreatitis mimicking a malignant solid tumour. BMJ Case Rep. 2015;2015(3):e717–50.

16. Heit D, Megson S, Cope-Yokoyama S, Goyal A. Pancreatic head tumor in an infant with new-onset jaundice. J Pediatr Gastroenterol Nutr. 2016;62(1):e14-5.

17. Hirose K, Nakamura T, Takahashi N, Shida Y, Hasuo K, Koizuka H. Xanthogranulomatous pancreatitis associated with intraductal papillary mucinous tumor. AJP Am J Roentgenol. 2005;183(4):707–13.

18. Iyer VK, Aggarwal S, Mathur M. Xanthogranulomatous pancreatitis: mass lesion of the pancreas simulating pancreatic carcinoma—a report of two cases. Indian J Pathol Microbiol. 2004;47(1):36–8.

19. Kamitani T, Nishimiya M, Takahashi N, Shida Y, Hasuo K, Koizuka H. Xanthogranulomatous pancreatitis associated with intraductal papillary mucinous tumor. AJP Am J Roentgenol. 2005;183(4):707–13.

20. Kato M, Kohyama T, Kojima T, Okamoto K, Ito S, Morita T, Araki K, Onishi S. Xanthogranulomatous pancreatitis assessed secondary to acute pancreatitis: two case reports. Hepatogastroenterology. 2007;54(78):1648–51.

21. Kim YN, Park SY, Kim YK, Moon WS. Xanthogranulomatous pancreatitis combined with intraductal papillary mucinous carcinoma in situ. J Korean Med Sci. 2010;25(12):1814–7.

22. Kim HS, Joo M, Chang SH, Song HY, Song TJ, Seo JW, Kim CN. Xanthogranulomatous pancreatitis presents as a solid tumor mass: a case report. J Korean Med Sci. 2011;26(4):583–6.

23. Koyama Y, Ozaki K, Fukui Y, et al. Xanthogranulomatous change appearing in the pancreas cyst wall. Pancreas. 1993;8(5):649–51.

24. Iyer VK, Aggarwal S, Mathur M. Xanthogranulomatous pancreatitis: mass lesion of the pancreas simulating pancreatic carcinoma—a report of two cases. Indian J Pathol Microbiol. 2004;47(1):36–8.

25. Kamitani T, Nishimiya M, Takahashi N, Shida Y, Hasuo K, Koizuka H. Xanthogranulomatous pancreatitis associated with intraductal papillary mucinous tumor. AJP Am J Roentgenol. 2005;183(4):707–13.

26. Okabayashi T, Nishimori I, Kobayashi M, Sugimoto T, Kohsaki T, Okamoto K, Ito S, Morita T, Araki K, Onishi S. Xanthogranulomatous pancreatitis assessed secondary to acute pancreatitis: two case reports. Hepatogastroenterology. 2007;54(78):1648–51.

27. Shimizu S, Saisaka Y, Furukita Y, Nishimura T, Homiri T, Nakamura T, Tanaka K, Shibuya Y, Ozaki K, Fukui Y, et al. Resected xanthogranulomatous pancreatitis. J Hepatobiliary Pancreat Surg. 2008;15(2):240–2.