Epidermoid cyst arising from an intrapancreatic accessory spleen: A case report and review of the literature

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Abstract. We describe an epidermoid cyst arising from an accessory spleen of the pancreas. A 56-year-old female with iron deficiency anemia presented with dizziness. During the clinical workup, a 2x4 cm-sized mass was incidentally detected in the tail of the pancreas in a computed tomography (CT) scan. Under a clinical diagnosis of pancreatic cystic malignant tumor, a distal pancreatectomy was carried out. Grossly, the lesion was composed of a solid and cystic portion. Microscopic analysis revealed that the solid portion was an intrapancreatic accessory spleen and the cystic portion was an epidermoid cyst. An epidermoid cyst in an intrapancreatic accessory spleen is extremely rare and hence difficult to diagnose pre-operatively. Taking into account the possibility of such a cyst in the differential diagnosis of intrapancreatic cystic lesion is recommended.

Introduction

The occurrence of an accessory spleen is not rare; it affects approximately 10% of the general population, and 16% of all cases are intrapancreatic (1). However, the development of an epidermoid cyst of intrapancreatic accessory spleen (IPAS) is not common, with 30 cases (2-29) described in the literature since Davidson et al (2) described the first case of epidermoid cyst of IPAS. Due to the difficulty in differentiating the lesion from a cystic neoplasm of the pancreas by an imaging study (4), the majority have been diagnosed following surgical resection, with the exception of one case by Itano et al (5). Pre-operative diagnosis was mainly cystic neoplasm of the pancreas. Herein, we report a case of 54-year-old female with an epidermoid cyst of an IPAS and review the literature.

Case report

A 54-year-old female with iron deficiency anemia was admitted to hospital complaining of dizziness and abdominal discomfort. During the clinical workup, a 2.3 cm-radiological-sized cystic mass was detected in the tail of the pancreas by abdominal computed tomography (CT; Fig. 1). Distal pancreatectomy was performed upon clinical diagnosis of pancreatic cancer. Grossly, the surgical specimen showed a well-demarcated multilocular cystic mass within the pancreatic parenchyma, measuring 2.0x1.5 cm (histological size) and containing dark serosanguineous fluid. Microscopic investigation revealed that the majority of the epithelial lining was comprised of multilayered cuboidal epithelium with focal denudation. However, no atypical or malignant changes were observed (Fig. 2). Immunohistochemical staining demonstrated that the epithelial lining was reactive for cytokeratin (CK) and CK7. The cystic wall demonstrated histologically normal splenic pulp tissue, which was surrounded by a hyalinized fibrous band. The final pathologic diagnosis was an epidermoid cyst arising from an IPAS. The six-month post-operative course was uneventful.

Discussion

Approximately 16% of accessory spleens occur in or around the tail of the pancreas (1). An epidermoid cyst in an IPAS is extremely rare and was first described in 1980 by Davidson et al (2). Following this, 30 cases of epidermoid cyst of IPAS have been described in the literature. Table I summarizes the 31 cases of epidermoid cyst in an IPAS, including the case we describe here. The cases involved 15 males and 16 females, with ages ranging from 12-70 years (mean, 46 years). All cysts were located in the pancreatic tail. While 16 patients were asymptomatic, various symptoms were observed in 14 patients, including weight loss, nausea, vomiting, abdominal pain and discomfort, back pain, epigastric pain and fever. Histological analysis revealed that the cysts were solitary or...
multilocular, lined with keratinized or non-keratinized stratified squamous epithelium or cuboidal epithelium, and in some cases exhibiting mixed-form epithelium.

An elevation of serum CA 19-9 level was observed in 10 cases, hence the difficulty in pre-operatively differentiating between an epidermoid cyst in an IPAS and pancreatic malignancy during clinical analysis. Higaki et al. (9) revealed that the serum CA 19-9 level markedly decreased to within the normal range following surgery in a patient diagnosed with an epidermoid cyst in an IPAS, suggesting that the serum CA 19-9 originated in the epidermoid cyst in an IPAS.

Table I. Summary of the 31 cases of epidermoid cyst arising in intrapancreatic accessory spleen (IPAS), including the present case.

| No | First author (Ref.) | Year | Age/No | Gender | Site | Symptoms | CA 19-9 | Size (cm) | Surgery | Epi. lining |
|----|----------------------|------|--------|--------|------|----------|---------|-----------|---------|------------|
| 1  | Davidson (2)         | 1980 | 40/M   | Tail   | WL, N | NR       | 5.5     | DP&S      | -       | -          |
| 2  | Hanada (3)           | 1981 | 51/M   | Tail   | AP    | NR       | -       | DP        | -       | -          |
| 3  | Jibu (4)             | 1987 | 37/M   | Tail   | -     | -        | 4.0     | -         | -       | -          |
| 4  | Morohoshi (5)        | 1991 | 32/M   | Tail   | AP    | WNL      | 6.0x5.0 | RC        | S        | SSE        |
| 5  | Naka (6)             | 1991 | 37/F   | Tail   | AP    | NR       | -       | SPDP      | -       | -          |
| 6  | Tang (7)             | 1994 | 38/M   | Tail   | ASx.  | NR       | 1.4     | DP        | SSE      |            |
| 7  | Furukawa (8)         | 1998 | 45/M   | Tail   | ASx.  | WNL      | 2.0     | DP        | SSE      |            |
| 8  | Higaki (9)           | 1998 | 46/F   | Tail   | Back pain | 201  | 3.0x3.0 | DP&S      | SSE      |            |
| 9  | Tateyama (10)        | 1998 | 67/F   | Tail   | AP    | 201      | 3.0     | DP        | SSE      |            |
| 10 | Sasou (11)           | 1999 | 49/F   | Tail   | ASx.  | WNL      | 4.3x2.6 | DP        | NSSE     |            |
| 11 | Tsutsumi (12)        | 2000 | 51/M   | Tail   | ASx.  | WNL      | 2.5     | DP        | NSSE     |            |
| 12 | Choi (13)            | 2000 | 54/F   | Tail   | EP, N, V, WL | NR   | 15x11.0 | E&S       | KSSE     |            |
| 13 | Horibe (14)          | 2001 | 48/M   | Tail   | ASx.  | 53       | 2.0x1.0 | DP        | SSE      |            |
| 14 | Sonomura (15)        | 2002 | 45/F   | Tail   | EP    | 159      | 3.5     | DP        | SSE      |            |
| 15 | Yokomizo (16)        | 2002 | 38/M   | Tail   | ASx.  | 410      | 2.7     | DP        | NSSE     |            |
| 16 | Fink (17)            | 2002 | 12/F   | Tail   | Fever | NR       | 10.0    | RC        | NSSE     |            |
| 17 | Kanazawa (18)        | 2004 | 58/F   | Tail   | ASx.  | 62       | 2.5     | SPDP      | SSE      |            |
| 18 | Ru (19)              | 2007 | 41/M   | Tail   | ASx.  | NR       | 2.5     | DP        | NSSE     |            |
| 19 | Itano (20)           | 2008 | 40/M   | Tail   | ASx.  | WNL      | 3.0     | DP        | SSE      |            |
| 20 | Servais (21)         | 2008 | 52/F   | Tail   | ASx.  | NR       | 11.5x10.5x8.5 | DP | CCE        |
| 21 | Gleeson (22)         | 2008 | 32/F   | Tail   | AP    | NR       | 1.5x1.2 | DP&S      | SSE      |            |
| 22 | Kadota (23)          | 2009 | 57/F   | Tail   | ASx.  | WNL      | 6.0x5.0x4.0 | DP | NSSE, CE  |
| 23 | Kadota (23)          | 2009 | 70/F   | Tail   | ASx.  | 48       | 1.7x1.0x0.8 | DP | NSSE, CE  |
| 24 | Kadota (23)          | 2009 | 37/M   | Tail   | ASx.  | 647      | 10x7x0.7 | SPDP      | KSSE, CE |
| 25 | Zhang (24)           | 2009 | 52/F   | Tail   | ASx.  | WN       | 2.5x2.5 | SPDP      | SSE      |            |
| 26 | Itano (25)           | 2010 | 67/M   | Tail   | EP, WL | WNL      | 3.0     | LAPD      | SSE      |            |
| 27 | Yamanishi (26)       | 2011 | 55/F   | Tail   | ASx.  | 90       | 2.5x1.5 | DP        | SSE      |            |
| 28 | Iwasaki (27)         | 2011 | 36/F   | Tail   | EP, WL | 79       | 3.4x1.9 | LDP       | SSE      |            |
| 29 | Horn (28)            | 2011 | 62/M   | Tail   | AP    | NR       | 4.8x3.7x1.9 | CR | KSSE      |
| 30 | Khashab (29)         | 2011 | 49/F   | Tail   | AP    | NR       | 2.3     | LAPD      | SSE      |            |
| 31 | Present case         | 2012 | 56/F   | Tail   | AP    | WNL      | 2.0x1.5 | SPDP      | CE       |            |

M, male; F, female; Epi. lining, epithelial lining; EP, epigastric pain; N, nausea; V, vomiting; WL, weight loss; ASx., asymptomatic; CA 19-9, carbohydrate antigen 19-9 (IU/ml); DP, distal pancreatectomy; LAPD, laparoscopic-assisted distal pancreatectomy; LDP, laparoscopic distal pancreatectomy; NR, not reported; RC, removal of the cyst; SPDP, spleen-preserving distal pancreatectomy; ES, explorolaparotomy; S, splenectomy; NSSE, non-keratinizing stratified squamous epithelium; CE, cuboidal epithelium; KSSE, keratinizing stratified squamous epithelium; SSE, stratified squamous epithelium; CCE, columnar and cuboidal epithelium.

The histogenesis of an epidermoid cyst in an IPAS may be identical to that of a splenic epidermoid cyst (23). There are three hypotheses concerning the histogenesis of an epidermoid cyst in an IPAS (10). Firstly, the cyst may originate from mesothelial inclusion with subsequent squamous metaplasia (30). Secondly, teratomatous derivation or an inclusion of fetal squamous epithelium may cause cystic change (31). Thirdly, a derivation from the pancreatic duct may protrude into the accessory spleen (10). In a case described by Kadota et al (23), there were pancreatic ducts in the fibrous tissue surrounding the accessory spleen tissue, and the squamous and cuboidal...
epithelia indicated a transitional appearance from one form to the other. Additionally, immunohistochemical analysis demonstrated that the staining results of the cystic epithelial lining were identical to those of the pancreatic duct. These results support the third hypothesis.

A pre-operative imaging diagnosis of an epidermoid cyst in an IPAS is extremely difficult. Notably, a diagnosis of abdominal CT in the present case was also pancreatic tail cancer. As there are no characteristic features to define the lesion on radiology, it is not possible to entirely differentiate the cystic pancreatic malignancy prior to surgery and histopathological examination (28).

In conclusion, an epidermoid cyst in an IPAS is an extremely rare disease entity, and radiographic and clinical results (including CA 19-9 elevation) are similar to those of other cystic pancreatic neoplasms. As a result, the possibility of such a cystic lesion should be considered in the differential diagnosis of a pancreatic cystic lesion.

Figure 1. Radiological examination. Enhanced abdominal computed tomography (CT) reveals a clearly defined, 2.3 cm-sized nodule in the pancreatic tail (arrow).

Figure 2. Histopathological examination. (A) Grossly, the cut surface of the pancreas exhibits a multilocular cystic mass measuring 2.0x1.5 cm. (B,C) Microscopic analysis reveals a multilocular cyst (circle) surrounded by accessory splenic tissue (star) in the pancreas parenchyma (triangle). (D) The cyst is lined by multilayered cuboidal epithelium (arrow).
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