bin 0.2 mg/dl, and indirect bilirubin 0.7 mg/dl. The metabolic diseases screening test results were: blood ammonium level 50 mmol/L (normal), reducing substance in the urine (negative), and tandem mass spectrometry (normal). Echocardiography was normal.

To our knowledge, fatty liver degeneration in Seckel syndrome has not been reported previously. There is considerable heterogeneity in the clinical characteristics in Seckel syndrome (5-7). As a result of this case report, we suggest also investigating patients for hepatosteatosis.

REFERENCES

1. Seckel HPG. Bird-headed dwarfs. Springfield, IL: Charles C Thomas, 1960; 241.
2. Can E, Bulbul A, Uslu S, et al. A case of Seckel syndrome with tetralogy of Fallot. Genet Couns 2010; 21: 49-51.
3. Alderton, GK, Joenje H, Varon R, et al. Seckel syndrome exhibits cellular features demonstrating defects in the ATR-signalling pathway. Hum Mol Genet 2004; 13: 3127-38.
4. Casper AM, Durkin SG, Arlt MF, Glover TW. Chromosomal instability at common fragile sites in Seckel syndrome. Am J Hum Genet 2004; 75: 654-60.
5. Faivre L, Le Merrer M, Lyonnet S, et al. Clinical and genetic heterogeneity of Seckel syndrome. Am J Med Genet 2002; 112: 379-83.
6. Takikawa KM, Kikuchi A, Yokoyama A, et al. Perinatal findings of Seckel syndrome: a case report of a fetus showing primordial dwarfism and severe microcephaly. Fetal Diagn Ther 2008; 24: 405-8.
7. Rahme R, Crevier L, Dubois J, et al. Moyamoya-like vasculopathy and Seckel syndrome: just a coincidence? Childs Nerv Syst 2010; 26: 983-6.

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An unusual case of polycythemia vera with a complication of pancreatic pseudocyst

Polistemia vera komplikasyonu olarak gelişen bir nadir pankreatik psödokist vakası

To the Editor,

Polycythemia vera (PV) is a myeloproliferative disorder that can be complicated with thrombosis. The rate of thrombosis is as high as 50% (1-7). Budd-Chiari syndrome and portal, splenic, or mesenteric vein thrombosis are some examples of the major thrombotic events that can be seen in PV patients. Portal hypertension and hypersplenism can complicate as a result (8,9).

Pancreatic pseudocyst is defined as a fluid collection >4 weeks old and surrounded by a defined wall (10). Most pseudocysts include sterile material. There is no need for treatment for an asymptomatic pseudocyst (11,12).

In this article, we report a case of PV with a rare secondary complication of pancreatitis.

A 46-year-old male had symptoms of abdominal pain, pruritus and dyspnea. During the previous four months, he had noted these complaints with increasing severity. Physical examination revealed hepatomegaly and splenomegaly. He had also rales in the left hemithorax.

On laboratory examination, hematocrit level was 57.02 x10^3/µl (37.0–50.0), leukocyte count 17.6 x10^3/µl (4.0–11.0), granulocyte count 17.0 x10^3/µl (9.0–17.0), and platelet count 734 x10^3/µl (150–400), and JAK2 mutation was present. These findings were consistent with PV. There were no signs of another clotting disorder. Phlebotomy and hydroxyurea therapy was started.

Computed tomography (CT) revealed portal vein

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thrombosis with cavernous transformation and collaterals. The CT showed normal appearance of the liver with a mild splenomegaly (14 cm). There were hypodense areas in the spleen compatible with infarction. Splenic artery thrombosis was observed in the CT. There was also a cystic appearance in the tail of the pancreas, with a cyst diameter of 8 cm. CT also showed free abdominal ascites. The left hemithorax was filled with massive effusion. There was a filling defect in the left distal pulmonary vein compatible with a pulmonary vein embolus. Upper endoscopy revealed grade 3 esophageal and fundic varices.

The endosonographic (EUS) examination revealed a pancreatic cyst with regular border. The maximal diameter of the cyst was 8 cm. There was an anechoic cystic appearance in the EUS. All these findings correlated with a pseudocystic view (Figure 1).

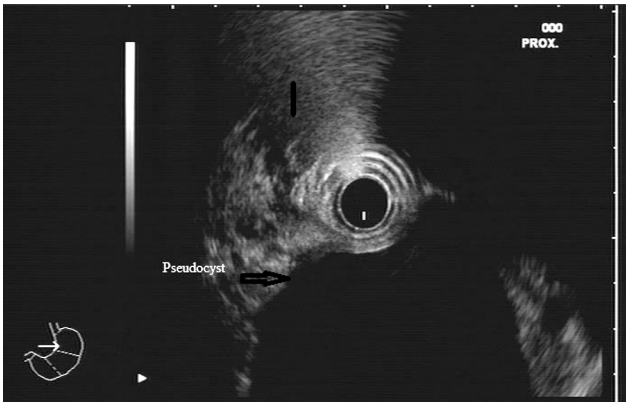

Figure 1. The endosonographic view of the patient with pancreatic pseudocyst as a result of thrombosis caused by polycythaemia vera

Low molecular weight heparin was started as a treatment for the thrombotic disorder. After discharge from the hospital, EUS examination was performed at three-month intervals for one year. The diameter of the pancreatic cyst was unchanged, and the patient was well over this time period.

Portal, splenic and mesenteric thrombosis can be observed frequently in PV patients (13). There is no clear explanation for the cause of thrombosis occurrence in these sites. It is supposed that hyperviscosity may play a significant role (14). It has been concluded that approximately 15% of PV patients have thrombotic complication before diagnosis (2).

Pancreatic pseudocysts occur in 2–10% of patients with mild and in approximately 50% of patients with severe acute pancreatitis (15). In the EUS, the pseudocyst appears with lack of septations and mural nodules; however, internal debris is seen frequently. Pseudocysts can be aspirated under EUS guidance for a differential diagnosis of mucinous cystic neoplasms (16,17). Because of the high bleeding risk and stability of the cyst size, we did not perform a drainage procedure in our case.

There are some reports regarding the benefit of splenectomy for PV patients with portal thrombosis. Randi et al. (18) found that increased use of cytotoxic drugs during the early postoperative period can increase the risk of leukemic transformation. They also stated that patients who survive the first post-splenectomy period may have a long and safe life (18). Because of the stability of our patient, we controlled our patient with hydroxyurea treatment. The pancreatic pseudocyst was asymptomatic during the follow-up of our patient. There was no other pancreatitis episode during the follow-up visits.

As mentioned above, thrombotic episodes can be seen in PV patients. To our knowledge, there has been no previous case in the literature with pancreatic pseudocyst as a complication of PV. At the time of this report, in spite of the simultaneous portal and pulmonary thrombosis episode, the patient was asymptomatic with hydroxyurea and anticoagulation treatment.

REFERENCES

1. Elliott MA, Tefferi A. Thrombosis and haemorrhage in polycythaemia vera and essential thrombocythaemia. Br J Haematol 2005; 128: 275-90.
2. Gruppo Italiano Studio Policitemia. Polycythemia vera: the natural history of 1213 patients followed for 20 years. Ann Intern Med 1995; 123: 656-64.
3. Marchioli R, Finazzi G, Landolfi R, et al. Vascular and neoplastic risk in a large cohort of patients with polycythemia vera. J Clin Oncol 2005; 23: 2224-32.
4. Cortelazzo S, Viero P, Finazzi G, et al. Incidence and risk factors for thrombotic complications in a historical cohort of 100 patients with essential thrombocythemia. J Clin Oncol 1990; 8: 556-62.
5. Besses C, Cervantes F, Pereira A, et al. Major vascular complications in essential thrombocythemia: a study of the predictive factors in a series of 148 patients. Leukemia 1999; 13: 150-4.

6. Passamonti F, Rumi E, Pungolino E, et al. Life expectancy and prognostic factors for survival in patients with polycythemia vera and essential thrombocythemia. Am J Med 2004; 117: 755-61.
7. Wolanskyj AP, Schwager SM, McClure RF, et al. Essential thrombocythemia beyond the first decade: life expectancy, long-term complication rates, and prognostic factors. Mayo Clin Proc 2006; 81: 159-66.
8. Acharya J, Westwood NB, Sawyer BM, et al. Identification of latent myeloproliferative disease in patients with Budd-Chiari syndrome using X-chromosome inactivation patterns and in vitro erythroid colony formation. Eur J Haematol 1995; 55: 315-21.
9. De Stefano V, Teofili L, Leone G, Michiels JJ. Spontaneous erythroid colony formation as the clue to an underlying myeloproliferative disorder in patients with Budd-Chiari syndrome or portal vein thrombosis. Semin Thromb Hemost 1997; 23: 411-8.
10. Bradley EL 3rd. A clinically based classification system for acute pancreatitis. Summary of the International Symposium on Acute Pancreatitis, Atlanta, Ga, September 11 through 13, 1992. Arch Surg 1993; 128: 586.
11. Vitas GJ, Sarr MG. Selected management of pancreatic pseudocysts: operative versus expectant management. Surgery 1992; 111: 123.
12. Yeo CJ, Bastidas JA, Lynch-Nyhan A, et al. The natural history of pancreatic pseudocysts documented by computed tomography. Surg Gynecol Obstet 1990; 170: 411.
13. Valla D, Benhamou JP. Obstruction of the hepatic veins or suprahepatic inferior vena cava. Dig Dis 1996; 14: 99-118.
14. Randi ML, Rossi C, Tison T, et al. Portal thrombosis and myeloproliferative diseases. Int Congress Phlebolymphology 1991; 21.1-21.3 [Abstract].
15. Wilson C. Management of the later complications of severe acute pancreatitis – pseudocyst, abscess and fistula. Eur J Gastroenterol Hepatol 1997; 9: 117-21.
16. Fockens P, Johnson TG, van Dullemen HM, et al. Endosonographic imaging of pancreatic pseudocysts before endoscopic transmural drainage. Gastrointest Endosc 1997; 46: 412-6.
17. Norton ID, Clain JE, Wiersema MJ, et al. Utility of endoscopic ultrasonography in endoscopic drainage of pancreatic pseudocysts in selected patients. Mayo Clin Proc 2001; 76: 794-8.
18. Randi ML, Fabris F, Ruzzon E, et al. Haematologica. Splenectomy after portal thrombosis in patients with polycythemia vera and essential thrombocythemia. Haematologica 2002; 87: 1180-4.

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