CASE REPORT

doi: 10.5455/medarh.2023.77.314-318
MED ARCH. 2023; 77(4): 314-318
RECEIVED: JUN 13, 2023
ACCEPTED: AUG 04, 2023

[1]Departments of General Surgery, King Fahd University Hospital, Dammam, Saudi Arabia

[2]College of Medicine, Imam Abdulrahamn Bin Faisal University, Dammam, Saudi Arabia

Corresponding author: Arwa Hanafie M. Ibrahim (MBBS). Department of General Surgery. King Fahd University Hospital, Dammam, Saudi Arabia. E-mail: arwa.ibrahim95@hotmail.com. ORCID ID: http://www.orcid.org/0000-0000-0000-0000-

Giant Sclerosing Hepatic Hemangioma Presenting as Bornman-Terblanche-Blumgart Syndrome: a Case Report and Review of the Literature

Arwa H. Ibrahim[1], Dhuha N. Boumarah[1], Arwa A. AlGhamdi[1], Shadi A. Alshammary[1,2]

ABSTRACT

Background: Hepatic hemangioma represents the most frequent benign tumor originating from the liver. When the tumor exceeds 10 cm, and in some studies 4 or 5 cm, it is considered giant, which accounts for 10% of all hemangiomas arising from the liver. Histologically, Sclerosing hepatic hemangioma, in particular, is an exceedingly rare subtype of hemangioma. Clinically Bornman-Terblanche-Blumgart syndrome is a very rare complication of hepatic hemangioma. **Objective:** The aim of this case presentation was to contribute to the literature by documenting a case of giant sclerosing hemangioma diagnosed in a 36-year-old female presenting with Bornman-Terblanche-Blumgart syndrome, along with a brief review of the literature. **Case report:** The current paper documents two rare clinical and histological features of hepatic hemangioma. Bornman-Terblanche-Blumgart syndrome is complicated a giant hepatic hemangioma found histologically to be sclerosing in nature. Knowledge about the uncommon complications of liver hemangioma permits the implementation of appropriate interventions in a timely manner and, in turn, can enhance the patient's quality of life and minimize rates of associated mortality.

Keywords: Giant Sclerosing Hepatic Hemangioma.

1. BACKGROUND

Hepatic hemangioma represents the most frequent benign tumor originating from the liver. It is formed by clusters of vascular malformations and drives its blood supply from the hepatic artery (1, 2). The prevalence of liver hemangioma has been estimated to reach 20% in autopsy studies (3). When the tumor exceeds 10 cm, and in some studies 4 or 5 cm, it is considered giant, accounting for 10% of all hemangiomas arising from the liver (2, 4, 5). These lesions are usually asymptomatic and present in a solitary pattern (5). Sclerosing hepatic hemangioma, in particular, is an exceedingly rare subtype of hemangioma characterized by the presence of degenerative changes histologically (6). To date, few reports have been published to comprehensively address the clinical features of this uncommon condition.

Bornman-Terblanche-Blumgart syndrome, a triad of abdominal pain and fever with unremarkable laboratory values, is a rare complication of hepatic hemangioma. These syndromic clinical features are recognized as a valid indicator of an ongoing inflammatory process within the hemangioma cavity (7). Although rare, the entity remains clinically significant as it possesses several diagnostic challenges and mandates surgical intervention (8). Herein, we contribute to the literature by documenting a case of giant sclerosing hemangioma diagnosed in a 36-year-old female presenting with Bornman-Terblanche-Blumgart syndrome, along with a brief review of the literature.

2. OBJECTIVE

The aim of this case presentation was to contribute to the literature by documenting a case of giant sclerosing hemangioma diagnosed in a 36-year-old female presenting with Bornman-Terblanche-Blumgart syndrome, along with a brief review of the literature. The present case was reported in accordance with the SCARE guidelines (9).

Author/ year	Age/ sex	Clinical presentation	Laboratory finding	Radiologic finding	Surgical approach	Intra-operative bleeding/ blood transfusion requirement	Post-operative complications
Lee C et al., 1994 [17]	37/ F	Fever	WBC: normal LFT: normal Inflammatory markers: N/ A	Rt. Posterior segment 5cm	Conservative	N/A	N/A
Khalid M et al., 2013 [18]	50/ M	Epigastric mass, fever, weight loss and weakness	WBC: normal LFT: normal Inflammatory markers: elevated	Giant LH involve Segment 2 and 3, other lesions involve 4b, and 7	Surgical resection of the mass	N/A	None
Hao F et al., 2017 [19]	52 / F	Fever	WBC: normal LFT: normal Inflammatory markers: elevated	Rt. Lobe Giant LH 16cm x 12cm	arterial embolization then Right hemihepatectomy	blood loss of 200 mL	None
Liu X et al., 2018 [20]	33/ M	Fever	WBC: normal LFT: normal Inflammatory markers: elevated	Rt. Lobe Giant LH 20cm	Right trisectionectomy	N/A	None
Dima-Cozma L et al., 2018 [21]	70/ F	Fever and abdominal pain	WBC: normal LFT: normal Inflammatory markers: elevated	7.3cm x 6.3cm LH involve Segment 4	segmentectomy	N/A	None
Pandit N et al., 2018 [22]	49/ F	Fever	WBC: normal LFT: normal Inflammatory markers: N/A	Left lateral segment Giant LH 15cm x 11cm	laparoscopic assisted left lateral segmentectomy	N/A	None
Desai G et al., 2020 [23]	38/ F	Fever, and abdominal discomfort	WBC: normal LFT: normal Inflammatory markers: elevated	Giant LH involve Segment 5, 6, 7, and 8	arterial embolization then Right hepatectomy	blood loss of 300 mL	None
Yoshimizu C et al., 2022 [24]	53/ F	Fever	WBC: normal LFT: normal Inflammatory markers: elevated	Giant LH involve Segment 8/7 and S3/2 10cm	Laparoscopic segmentectomy for both hemangioma	N/A	None

Table 1. Previously Reported Cases That Met the Diagnostic Criteria Of Bornman-Terblanche-Blumgart Syndrome

3. CASE PRESENTATION

We report a case of a 36-year-old Asian female known case of recurrent urolithiasis. She presented to the clinic after liver hemangioma was incidentally diagnosed in abdominal computed tomography (CT) scans of the abdomen performed for urolithiasis. The patient reported long-standing right upper quadrant and epigastric abdominal pain, on-and-off in nature, associated with nausea, vomiting, and early satiety. Her family history was unremarkable. At that time, CT showed a right lobe lesion measuring 14.6 x 15.7 x 13 cm, with peripheral discontinuous nodular enhancement and gradual central filling, with an area of low attenuation. as the lesion replaced the right lobe, the right hepatic vein was completely compressed. In addition, the right portal vein has reduced caliber (Figure 1A and 1B).

Tumor markers, including CEA, AFP, and CA19-9, were negative. Other laboratory indicators were unremarkable, except for slightly elevated liver enzymes; as a result, the decision to proceed with the surgical intervention was made. During the preoperative workup period, she developed unexplained fever spikes with negative biochemical or radiological sources for sepsis other than a liver hemangioma. The patient underwent a right hepatectomy with access to the abdominal cavity through a right-sided inverted L incision. The intraoperative ultrasound (US) was used to assess hemangioma and its relation to the middle hepatic vein and other major liver vasculature and bile ducts. Due to the large-sized hemangioma and mobilization difficulty, the right Glisson pedicle was approached first using Takasaki extrahepatic glissonian approach to allow shrinkage of the hemangioma. Infra hepatic IVC was then controlled to lower the venous return during transection and minimize back bleeding and pringle's maneuver. The hanging technique was applied to facilitate the transection. Despite the rupture of the hemangioma capsule, bleeding was managed by hemangioma resection. Hemostasis was confirmed at the end of the procedure and bile leak was ruled out using intraoperative methylene blue injection through the cystic duct. The operative timing was 240 minutes, and the estimated blood loss of 800 ml, Total Pringle time was 45 minutes (15 minutes clamped and 5 off-clamped). No inotropes were required intraoperatively, and the patient extubated well at the end of the procedure. The pathological result came as sclerosing hemangioma.

Follow-up CT angiography and duplex studies showed good contrast filling in the portal, middle, and left hepatic vein with good liver regeneration. In addition, duplex studies confirmed normal portal and hepatic artery and vein flow. However, despite patent inflow and outflow vasculature and good remnant liver, she developed a transient increase in her bilirubin levels and INR suggestive of post-hepatectomy liver failure (PHLF). She also developed ascites managed using abdominal drains and

Patient related factors	Age >65, comorbidities: Diabetes mellitus, Obesity, Hyperbilirubinemia, Renal insufficiency, Cardiopulmonary compromise and Thrombocytopenia
Liver related factors	Hepatic steatosis, Liver damage due to Chemotherapeutic agents, Cirrhosis and chronic liver disease
Operation related factors	Intraoperative blood loss and Blood transfusions (>1200 ml) Dissection techniques; vascular resections or IVC repair excessive dissection of tissues around the portal triad and hepatoduodenal ligament Remnant liver volume <25% Operating time > 240 min. Prolonged application of Pringle or TVE (total vascular exclusion) maneuver

Table 2. Risk factor for PHLF

diuretics. The bilirubin levels and INR normalized within one week of the operation with supportive treatment, but she required diuretics for three months post-discharge.

Postoperative follow-up imaging studies (at 3 and 6 months) showed well regeneration of the remnant liver with no signs of portal hypertension or ascites. In 12 months follow-up patient was in good condition, with no active complaint with normalization of liver enzyme.

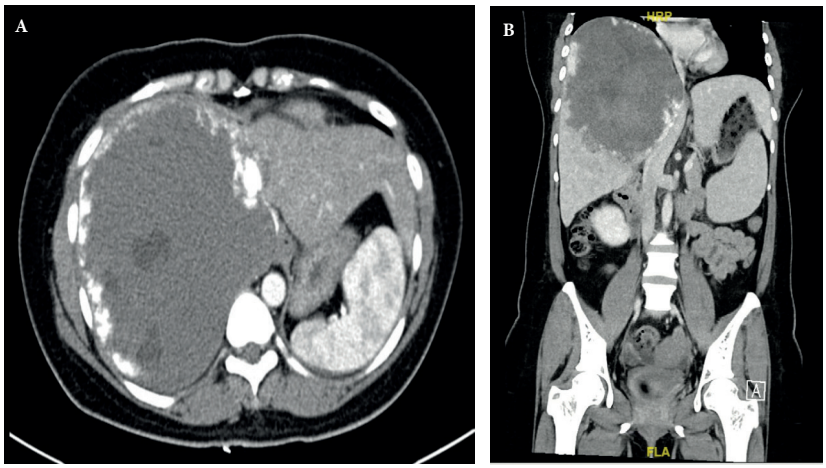

Figure 1. Axial (A) and Coronal (B) large liver lesion with discontinuous, nodular, peripheral enhancement in arterial phase, centripetal fill-in delayed portal venous phase confirming the diagnosis of giant hemangioma

4. DISCUSSION

Sclerosing hemangioma was first reported in 1983 by Shepherd and Lee in a series of four patients (10). Unlike typical hemangiomas, a slight male preponderance exists among reported cases, with the vast majority being in their sixties or seventies (11). However, our patient was a 36-year-old female who does not fit within the suggested demographic parameters. Localized deposition of hemosiderin particles, necrosis, thrombosis and/or hemorrhage can potentially induce the process of sclerosis in hepatic hemangioma. Abdominal pain or discomfort is the most commonly experienced symptom, secondary to Glisson's capsule stretching or compression on adjacent organs. Other symptoms, such as; nausea, vomiting, early satiety, and/or occasional episodes of fever have been reported (12, 13).

Nevertheless, most patients are asymptomatic and diagnosed incidentally during imaging for other reasons, as observed with our patient. Bornman-Terblanche-Blumgart syndrome represents a combination of abdominal pain and fever, with unremarkable laboratory markers and hepatic parameters seen among patients diagnosed with hepatic hemangioma (14-16). These clinical manifestations, despite the normal laboratory values, reflect an underlying inflammatory reaction within the hemangioma structure and the necessity of surgical treatment (7, 8, 16) The syndrome was named according to Bornman PC, Terblanche J, Blumgart RL, et al., who first described its clinical features. The authors reported their observations among four patients who were managed by successful surgical resection (16). The condition is recognized as a rare clinical entity reported only among a minority of patients (Table 1).

Ultrasonography, CT, and magnetic resonance imaging (MRI) are widely used imaging modalities. A homogeneous, hyperechoic, well-defined lesion that may exhibit posterior acoustic enhancement can present in the US. While both CT and MRI with contrast present, hypoattenuating mass, early peripheral contrast enhancement with a nodular pattern, progressive opacification, and isodense (centripetal) fill, depending on the study phase (11, 15). A typical hemangioma diagnosis can be reached with more than 90% sensitivity and specificity when MRI is utilized. In contrast, atypical morphology of hemangioma due to intralesional thrombosis, calcification, hyalinization, or cystic components can mimic malignant tumors that result in challenging diagnosis and necessitate histopathological correlation (25). The role of a biopsy in liver hemangioma is less significant, considering the benign nature and the risk of bleeding after the biopsy. However, some researchers advocate for biopsy to avoid unnecessary surgery (13, 26, 27). Based on the hypothesis that preoperative fluorodeoxyglucose positron emission tomography (FDG-PET) use could be helpful in the diagnosis as it distinguishes benign sclerosed hemangioma from malignant tumors, song, and shin reported iso- to a slightly decreased metabolic activity of the hemangioma on an FDG-PET scan (11, 25, 27).

The management of liver hemangioma can be one of three approaches, observational, non-surgical management, or surgical intervention. Asymptomatic, non-complicated hemangioma are safely observed and followed radiologically (27). At the same time, surgical

intervention is indicated for symptomatic patients or those who develop complications during follow-ups, such as coagulation disorders (Kasabach-Merritt syndrome) or Bornman-Terblanche-Blumgart syndrome, as seen with our patient (12). Nevertheless, the size of the hemangioma is not an indication (28). Both transarterial embolization (TAE) and radiofrequency ablation (RFA) are used for management and preoperative preparation (28). TEA is used to reduce symptoms, stabilize ruptured lesion preoperatively, and decrease lesional blood supply; however, recurrence is commonly observed, with no significant change in tumor size (3, 29). RFA, on the other hand, was found to have the ability to reduce the size of hepatic lesions (13, 28).

Surgery is the most effective therapeutic modality; four procedures can be applied: resection, enucleation, hepatic artery ligation, and transplantation (3, 28). The choice of procedure depends on location, number, and size of the tumor, presence of complications, surgeon preference, and experience (28). Both resection and enucleation are widely used, keeping transplantation as a last resort for diffuse, unresectable hepatic hemangiomatosis (30). The controversy between resection and enucleation arose from the fact that hemangioma is a benign disease, and removal of the normal liver parenchyma should be avoided. Several authors preferred enucleation over resection as it is associated with less intraoperative bleeding, risk of bile duct injury, shorter operative time, lower morbidity, shorter hospital stay, and maximum preservation of functional liver parenchyma. However, these advantages were observed in studies with an average hemangioma size of less than 10 cm. Few reports have described haemangiomas ≥ 10 cm and concluded with no statistically significant difference in both procedures in terms of surgical time, blood loss, and overall complications /12, 13, 28, 31). Minimally invasive (laparoscopic resection) can also be applied, especially on anteriorly located hemangioma (32, 33). Based on the literature, many complications secondary to surgical intervention on giant liver hemangioma have been reported, such as bleeding, pleural effusion, abdominal abscesses, biliary fistula or leakage, and mortality. Significant blood loss intra- or postoperative is a commonly encountered event (12). Dong et al. and Zhang et al. concluded in their studies that hemangioma size is an independent risk factor for complications (12, 31).

Although attempting to minimize intraoperative blood loss using early clamping of the right Glisson pedicle, intermittent pringle clamping, and partial infra hepatic IVC clamping, the blood loss was high secondary to a ruptured capsule of the hemangioma. High CVP pressure and over-resuscitation are associated with increased intraoperative blood loss in liver surgery (34).

Based on the literature, the incidence of post-hepatectomy liver failure (PHLF) ranges between 0.7-35%, with variation according to the preoperative status of the liver and underlying pathology. The risk factors are classified into patient, liver, and operation related. (Table 2). In our case, the main factor was operation-related blood loss with transfusion and dissection close to IVC and porta hepatis (35, 36).

Several studies showed no increased risk of major complications following liver resection in relation to hospital volume. But better communication between the caring teams and the surgeons, and the presence of a specialized anesthetist and intensivist to care for major liver patients are associated with improved outcomes and results.

5. CONCLUSION

Bornman-Terblanche-Blumgart syndrome is one of the clinical complications of a hepatic hemangioma that has been rarely addressed in the literature. The current paper documents the occurrence of two rare clinical and histological features of hepatic hemangioma in which Bornman-Terblanche-Blumgart syndrome complicated a giant hepatic hemangioma that was found histologically to be sclerosing in nature. Knowledge about the uncommon complications of liver hemangioma permits the implementation of appropriate interventions in a timely manner and, in turn, can enhance the patient's quality of life and minimize rates of associated mortality.

- **Author's contribution:** The all authors were involved in all steps of preparation this article. Final proofreading was made ny the first author.
- **Financial support and sponsorship:** None.
- **Conflicts of interest:** None.

REFERENCES

1. Bajenaru N, Balaban V, Săvulescu F, Campeanu I, Patrascu T. Hepatic hemangioma. J Med Life Sci. 2015; 8: 4–11.
2. Liu Y, Wei X, Wang K, Shan Q, Dai H, Xie H, et al. Enucleation versus anatomic resection for Giant Hepatic Hemangioma: A meta-analysis. Gastrointest Tumors. 2016; 3: 153–162.
3. Makal GB, Sonbahar BÇ, Özalp N. Surgical Treatment of Giant Liver Hemangioma, Case Report and Literature Review. Sisli Etfal Hastan Tip Bul. 2019; 53(3): 318-321.
4. Kaman L, Naik A, Savlania A, Raypattanaik N. Surgical Management of Giant Hepatic Haemangioma–Need for Redefining the Nomenclature According to the Size. Pol Przegl Chir. 2021 Mar 22; 93(4): 28-34.
5. Xie QS, Chen ZX, Zhao YJ, Gu H, Geng XP, Liu FB. Outcomes of surgery for giant hepatic hemangioma. BMC Surg. 2021 Apr 8; 21(1): 186.
6. Sugo H, Sekine Y, Miyano S, Watanobe I, Machida M, Kojima K, Okubo H, Ura A, Ogura K, Matsumoto T. Hepatic Sclerosing Hemangioma with Predominance of the Sclerosed Area Mimicking a Biliary Cystadenocarcinoma. Case Reports Hepatol. 2018 Oct 4; 2018: 7353170.
7. Duxbury MS, Garden OJ. Giant haemangioma of the liver: observation or resection? Dig Surg. 2010; 27(1): 7-11.
8. Ehrl D, Rothaug K, Herzog P, Hofer B, Rau HG. "Incidentaloma" of the liver: management of a diagnostic and therapeutic dilemma. HPB Surg. 2012; 2012: 891787.
9. Agha RA, Franchi T, Sohrabi C, Mathew G, for the SCARE Group. The SCARE 2020 Guideline: Updating Consensus Surgical CAse REport (SCARE) Guidelines, International Journal of Surgery 2020; 84: 226-230.

10. Shepherd NA, Lee G. Solitary necrotic nodules of the liver simulating hepatic metastases. J Clin Pathol. 1983 Oct; 36(10): 1181-1183.
11. Song JS, Kim YN, Moon WS. A sclerosing hemangioma of the liver. Clin Mol Hepatol. 2013 Dec; 19(4): 426-430.
12. Zhang W, Huang ZY, Ke CS, Wu C, Zhang ZW, Zhang BX, Chen YF, Zhang WG, Zhu P, Chen XP. Surgical Treatment of Giant Liver Hemangioma Larger Than 10cm: A Single Center's Experience With 86 Patients. Medicine (Baltimore). 2015; 94(34).
13. Hoekstra LT, Bieze M, Erdogan D, Roelofs JJ, Beuers UH, van Gulik TM. Management of giant liver hemangiomas: an update. Expert Rev Gastroenterol Hepatol. 2013 Mar; 7(3): 263-268.
14. Papafragkakis H, Moehlen M, Garcia-Buitrago MT, Madrazo B, Island E, Martin P. A case of a ruptured sclerosing liver hemangioma. Int J Hepatol. 2011; 2011: 942360.
15. Assy N, Nasser G, Djibre A, Beniashvili Z, Elias S, Zidan J. Characteristics of common solid liver lesions and recommendations for diagnostic workup. World J Gastroenterol. 2009 Jul 14; 15(26): 3217-3227.
16. Bornman original paper
17. Lee CW, Chung YH, Lee GC, Kim JY, Lee JS. A case of giant hemangioma of the liver presenting with fever of unknown origin. J Korean Med Sci. 1994 Apr; 9(2): 200-204.
18. Khalid M, Ahmad M, Jain A, Rizvi I. Atypical giant haemangioma of liver with systemic inflammatory manifestations. BMJ Case Rep. 2013 Jan 25.
19. Hao F, Yang X, Tian Y, Wang W, Ge M. Spontaneous internal hemorrhage of a giant hepatic hemangioma: A case report. Medicine (Baltimore). 2017 Nov; 96(47)
20. Liu X, Yang Z, Tan H, Zhou W, Su Y. Fever of Unknown Origin Caused by Giant Hepatic Hemangioma. J Gastrointest Surg. 2018 Feb; 22(2): 366-367.
21. Dima-Cozma LC, Bitere OR, Pantazescu AN, Gologan E, Mitu F, Rădulescu D, Cozma RS. Cavernous liver hemangioma complicated with spontaneous intratumoral hemorrhage: a case report and literature review. Rom J Morphol Embryol. 2018; 59(2): 557-561.
22. Pandit N, Awale L, Chaudhary S, Jaiswal LS. Fever of unknown origin: a rare presentation of giant hepatic hemangioma. J Surg Case Rep. 2018 Jun 14; 2018(6)
23. Desai G, Budkule D, Pande P, Wagle P. Pyrexia of Unknown Origin: An Atypical Presentation of Hepatic Hemangioma. Surg J (N Y). 2020 Dec 14;6(4)
24. Yoshimizu C, Ariizumi S, Kogiso T, Sagawa T, Taniai M, Honda G, Egawa H, Tokushige K. Giant Hepatic Hemangioma Causing Prolonged Fever and Indicated for Resection. Intern Med. 2022 Jun 15; 61(12): 1849-1856.
25. Shin YM. Sclerosing hemangioma in the liver. Korean J Hepatol. 2011 Sep; 17(3): 242-246.
26. Behbahani S, Hoffmann JC, Stonebridge R, Mahboob S. Clinical case report: Sclerosing hemangioma of the liver, a rare but great mimicker. Radiol Case Rep. 2016 May 6; 11(2): 58-61.
27. Li T, Klar MM, Alawad M, Abdul R, Zahiruddin A, Salifu MO, McFarlane IM. Hepatic Sclerosing Hemangioma Mimicking Malignancy: A Case and Literature Review. Am J Med Case Rep. 2021; 9(3): 144-146.
28. Boukerrouche A. Therapeutic options of giant liver hemangioma. J Liver Res Disord Ther. 2018; 4(1): 32-34.
29. Akamatsu N, Sugawara Y, Komagome M, Ishida T, Shin N, Cho N, et al. Giant liver hemangioma resected by trisectorectomy after efficient volume reduction by Transcatheter Arterial Embolization: A case report. J Med Case Rep. 2010; 4(1).
30. Ostojic A, Mrzljak A, Mikulic D. Liver transplantation for benign liver tumors. World J Hepatol. 2021 Sep 27; 13(9): 1098-1106.
31. Dong Z, Fang K, Sui C, Guo J, Dai B, Geng L, et al. The Surgical Outcomes and risk factors of giant hepatic haemangiomas: A single centre experience. Research Square. 2022.
32. Lanthaler M, Freund M, Nehoda H. Laparoscopic resection of a giant liver hemangioma. J Laparoendosc Adv Surg Tech A. 2005; 15(6): 624–626.
33. Gadiyaram S, Shetty N. Laparoscopic resection of giant liver hemangioma using laparoscopic Habib probe for parenchymal transection. J Minim Access Surg. 2012; 8(2): 59.
34. Melendez JA, Arslan V, Fischer ME, Wuest D, Jarnagin WR, Fong Y, Blumgart LH. Perioperative outcomes of major hepatic resections under low central venous pressure anesthesia: blood loss, blood transfusion, and the risk of postoperative renal dysfunction. J Am Coll Surg. 1998 Dec; 187(6): 620-625.
35. Ray S, Mehta NN, Golhar A, Nundy S. Post hepatectomy liver failure–A comprehensive review of current concepts and controversies. Ann Med Surg (Lond). 2018 Aug 23; 34: 4-10.
36. Ocak İ, Topaloğlu S, Acarli K. Posthepatectomy liver failure. Turk J Med Sci. 2020 Oct 22; 50(6): 1491-1503.