Desmoid Tumor of Rectus Abdominis Presenting with Grey-Turner’s and Cullen’s Sign: A Report of a Rare Case

Angoori Gnaneshwar Rao, T Swathi, Saba Syeda Farheen, Amit Kolli, Sharanya Hari, Uday Deshmukh Reddy, Kondapi Deepak, Kranthi Jagadevapuram

Abstract

Desmoid tumor of rectus abdominis presenting with Grey-Turner’s and Cullen’s sign is rare. Herein, we report desmoid tumor of rectus abdominis in a 64-year-old multiparous female who presented with ecchymosis involving left flank and around the umbilicus. Histopathological examination of biopsy from the tumor confirmed the diagnosis of the desmoid tumor. She was referred to a surgeon for radical resection.

Key Words: Cullen’s sign, desmoid tumor, Grey-Turner’s sign, radical resection

Introduction

Desmoid tumors are uncommon fibrous neoplasms originating from the musculoaponeurotic structures, account for 0.03% of all tumors and 3% of all soft-tissue neoplasms. The term desmoid was coined by Muller in 1838 and is derived from the Greek word desmos, which means tendon like.

Grey-Turner’s sign refers to bruising of the area between the last rib and top of the hip and is a sign of retroperitoneal hemorrhage. Cullen’s sign is superficial edema and bruising in the subcutaneous fatty tissue around the umbilicus. Causes include acute pancreatitis, ruptured ectopic, aortic rupture, and coagulopathies.

Case Report

A 64-year-old female presented with pain in the left flank for 2 months and bluish discoloration around the umbilicus and left flank of 10-day duration. There was no history of injury or intake of anticoagulants or personal or family history of bleeding diathesis. Examination revealed ecchymosis on the left flank extending onto the front of the abdomen and above the hip (Grey-Turner’s sign) and umbilicus and suprapubic region (Cullen’s sign). An ill-defined firm, tender swelling of 5 cm × 6 cm was palpable in the left parambilical region. The swelling became less prominent on leg raising test. Routine hematological investigations were unremarkable. Blood chemistry was within normal limits, and human immune deficiency virus serology was nonreactive. Abdominal computed tomography showed single well-defined oval intermediate signal intensity lesion arising from the rectus sheath on the left side. Fine-needle aspiration cytology from swelling showed the clusters of spindle cells with moderate pleomorphism, suggestive of desmoid tumor. Histopathology of biopsy from swelling showed the spindle-shaped cells in fasciculated bundles, most of them with a spindle to oval nuclei, confirmative of desmoid tumor. She was referred to a surgeon for radical surgical excision.

Discussion

As per the World Health Organization, desmoid tumors are defined as “clonal fibroblastic proliferations that arise in the deep soft tissues and are characterized by infiltrative growth and a tendency toward local invasion.”

From the Department of Dermatology, SVS Medical College, Mahbubnagar, Telangana, India

Address for correspondence:
Prof. Angoori Gnaneshwar Rao,
F12, B8, HIG-2 APHB,
Baghlingampally,
Hyderabad - 500 044, Telangana,
India.
E-mail: dr_a_g_rao@yahoo.co.in

What was known?
Certain clinical conditions such as acute pancreatitis, ruptured ectopic, aortic rupture, and coagulopathies are known to manifest with Grey-Turner’s and Cullen’s sign.
recurrence but an inability to metastasize”. Desmoids are characterized by mutations in the β-catenin gene, CTNNB1, or the adenomatous polyposis coli gene. The exact etiology of the desmoid tumor is not known; however, it has been postulated that the immunologic and hormonal changes in the pregnancy or postpartum play an important role. In addition, the local effect of the mechanical stress from a gravid uterus may also contribute to the occurrence of a desmoid tumor.[5]

It is prevalent among women of reproductive age group commonly occurring between the ages of 25 and 60 years. The abdominal desmoid tumor usually presents with pain; however, presentation of a desmoid tumor with Grey-Turner’s sign and Cullen’s sign in the index case is unique and rare. The anterior abdominal wall is the most common site of predilection with an incidence of 50%. Similarly, the index case also found to have a desmoid tumor in the anterior abdominal wall. Most abdominal wall desmoids measure between 5 cm and 15 cm in size. The size of desmoid (5/6 cm) in the index case is in concert with the size of most abdominal desmoids. Differential diagnosis of desmoid includes hypertrophic scars, nodular fasciitis, and fibroblastic sarcoma. Desmoid tumors are classified into three groups: within the abdominal wall, intra-abdominal, and extra-abdominal accounting for 25%, 15%, and 65%, respectively.[3] Extra-abdominal desmoid tumors are commonly found in the region of the shoulder girdle, trunk, and lower extremities. Furthermore, desmoid tumors are also known to occur in 10%–15% of patients with familial adenomatous polyposis. The association of intra-abdominal desmoids with familial adenomatous polyposis is known as Gardner’s syndrome. However, there was no association with familial adenomatous polyposis in the index case.

Radical resection is the ideal and most effective treatment for small and accessible desmoid tumors such as the index case.[4] However, big desmoid tumors require full thickness surgery and reconstruction with synthetic material. Recurrence occurs in up to 45% of patients which depends on the tumor size and on the resection.[5] Radiation therapy can be used for recurrent disease or as a primary treatment to avoid mutilating surgical resection. Moreover, it may be used preoperatively, postoperatively, or as the only treatment.[6] Various therapeutic agents have been tried in the management of desmoid tumor which includes Tamoxifen, anti-estrogen and Indomethcin, COX-2 inhibitor.[7-9]

Desmoid tumor presenting with Grey-Turner’s sign and Cullen’s sign has not been reported in the literature so far, and this may be the first case report.

In conclusion, a desmoid tumor of rectus abdominis should strongly be suspected in patients presenting with
Grey-Turner’s sign and Cullen’s sign. This may be added to the list of existing causes of Grey-Turner’s sign and Cullen’s sign.

**Financial support and sponsorship**
Nil.

**Conflicts of interest**
There are no conflicts of interest.

**What is new?**
Desmoid tumor of rectus abdominis presenting with Grey-Turner’s and Cullen’s sign is unique.

**References**
1. Economou A, Pitta X, Andreadis E, Papapavlou L, Chrissidis T. Desmoid tumor of the abdominal wall: A case report. J Med Case Rep 2011;5:326.
2. Johner A, Tiwari P, Zetler P, Wiseman SM. Abdominal wall desmoid tumors associated with pregnancy: Current concepts. Expert Rev Anticancer Ther 2009;9:1675-82.
3. De Cian F, Delay E, Rudigoz RC, Ranchère D, Rivoire M. Desmoid tumor arising in a cesarean section scar during pregnancy: Monitoring and management. Gynecol Oncol 1999;75:145-8.
4. Rohrich RJ, Lowe JB, Hackney FL, Bowman JL, Hobar PC. An algorithm for abdominal wall reconstruction. Plast Reconstr Surg 2000;105:202-16.
5. Nuyttens JJ, Rust PF, Thomas CR Jr., Turrisi AT 3rd. Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors: A comparative review of 22 articles. Cancer 2000;88:1517-23.
6. El-Haddad M, El-Sebaie M, Ahmad R, Khalil E, Shahin M, Pant R, et al. Treatment of aggressive fibromatosis: The experience of a single institution. Clin Oncol (R Coll Radiol) 2009;21:775-80.
7. Hansmann A, Adolph C, Vogel T, Unger A, Moeslein G. High-dose tamoxifen and sulindac as first-line treatment for desmoid tumors. Cancer 2004;100:612-20.
8. Waddell WR, Gerner RE. Indomethacin and ascorbate inhibit desmoid tumors. J Surg Oncol 1980;15:85-90.
9. Signoroni S, Frattini M, Negri T, Pastore E, Tamborini E, Casieri P, et al. Cyclooxygenase-2 and platelet-derived growth factor receptors as potential targets in treating aggressive fibromatosis. Clin Cancer Res 2007;13:5034-40.