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# Primary Omental Fibromatosis presenting as an Incarcerated Inguinal Hernia – Case Reports from a Single Institution over 20 years

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Abstract

INTRODUCTION:

Inguinal omental fibromatosis is a rare disease entity that may mimic incarcerated inguinal hernia clinically. We therefore review the incidence of inguinal omental fibromatosis in our center.

METHOD:

From 1.1.1996 to 30.6.2016, all hernia operations performed in all the affiliated Hospitals of the University of Hong Kong were reviewed retrospectively; data were retrieved from patient record inside clinical computer system.

RESULTS:

A total of 7039 hernia operations were carried out during the period in which 564 were incarcerated or strangulated hernia operations, among which, 2 cases were of diagnosis of omental fibromatosis, which account for incidence of 0.028% of groin exploration.

CASE REPORT:

This was second case in our center - A 26-year-old man was admitted with a history of reducible right groin mass since he was born, the mass had become irreducible for two months. Besides, the patient had no symptoms of bowel obstruction. On palpation, a firm mass was found in the right groin extending to the right scrotum, and could not be reduced completely. Bilateral testes in the scrotum were palpable. Computed

tomography scan of pelvic cavity showed that there was herniated omentum entered the right scrotum. The omental mass was resected completely and free-tension repair was performed. The histopathological examination revealed that the tumor consisted of spindle-shaped cells that consistent with fibromatosis.

Conclusion:

Inguinal omental fibromatosis is rare and it could present as incarcerated hernia clinically. Fibromatosis may be part of presentation of syndromal disease like Garden's syndrome, the recurrence is higher than in sporadic cases despite radical surgery.

## INTRODUCTION:

Fibromatosis is neoplastic, monoclonal myofibroblastic proliferations, may infiltrate the surrounding tissue but rarely metastasize [1]. The most common form of intra-abdominal fibromatosis is mesenteric fibromatosis [2, 3]. The intra-abdominal fibromatosis may remain asymptomatic, or cause complications according to their size and location of involvement, such as causing bowel obstruction, hydronephrosis, intestinal fistula, etc. The differential diagnosis includes gastrointesinal stromal tumors(GISTs), liposarcoma, lymphoma and other forms of inflammatory mass. Abdominal fibromatosis is associated with other systemic diseases like Crohn’s disease, or familial adenomatous polyposis syndrome, or Gardner syndrome. In patients with Garden's syndrome and abdominal fibromatosis, the chance of recurrence of fibromatosis after radical surgery is higher than in sporadic cases [4] .

## METHOD:

This is a retrospective study for incidence and clinical presentation of fibromatosis presented as inguinal mass in our center. From 1.1.1996 to 30.6.2016, operative diagnosis and procedure names of all inguinal hernia operations performed in three of the affiliated Hospitals of the University of Hong Kong – Queen Mary Hospital, HKSAR, China; Tung Wah Hospital, HKSAR, China and the University of Hong Kong – Shenzhen Hospital, Shenzhen China were retrieved from clinical management system

of Hospital Authority of Hong West Cluster, HKSAR, China and the University of Hong Kong – Shenzhen Hospital, Shenzhen, China were reviewed retrospectively.

**RESULTS:**

A total of 7039 hernia operations were carried out during the period in our centers as mentioned, in which 564 were either incarcerated or strangulated hernia operations, and 2 cases were of diagnosis of omental fibromatosis, which account for incidence of 0.028% of groin exploration, with our first case has already been reported in 1999 [5].

**ETHICS:**

Written consent was obtained from patient for reporting clinical presentation and progress of his disease for education and research purposes, with protection of patient's identity being non-identifiable.

**CASE REPORT:**

A 26-year-old man who enjoyed good past health presented as reducible right groin mass since childhood, size of inguinal mass increased recently and became irreducible for two months before admission. Apart from the inguinal mass, the patient did not suffer any symptoms of bowel obstruction. On physical examination, a 5cm non-tender irreducible firm mass was found in the right groin with inguino-scrotal extension. Trans-illumination test was negative and we could not get above the mass. Bilateral

testes were palpable in scrotum. With the given information, preliminary clinical diagnosis of irreducible inguinal hernia was made, with differential diagnosis of inguinal tumor, therefore imaging was performed to obtain a definitive diagnosis before proceeding to groin exploration. Computer Tomography showed that the right inguinal canal was dilated with the herniated omentum entered the right scrotum, and forming a tumor presented as a round soft tissue nodule (Figure 1 & 2). There was mild and homogeneous enhancement of the nodule after contrast enhancement was seen. Bilateral testes were normal. The differential diagnosis included: gastro-intestinal stromal tumor (GIST), inflammatory mass, or other malignant tumors. In view of uncertain nature of inguinal mass and inguinal hernia with possible incarceration, open right groin exploration was performed after discussing the pros and cons of surgery with patient.

Standardized right inguinal exploration was performed with skin incision, Scarpal fascia opened with inguinal canal was exposed. Anterior wall of inguinal canal was splitted with sharp cut and inguinal canal was explored: an indirect inguinal sac was identified and omentum tissue was seen extending from the deep inguinal ring, it wrapped around itself forming firm tumor in the hernia sac. The tumor was reduced completely from the indirect hernia sac which extended to scrotum after division of adhesion bands at the distal part of sac. The mass and the surrounding omentum was resected en-bloc, and Lichtenstein tension-free mesh repair was performed. The mass measured 5 × 4× 4cm, and the cut surface had a yellowish solid appearance (Figure 3).

The patient recovered uneventfully and was discharged 2 days after operation without any complication.

Histopathological examination revealed that the tumor consisted of spindle-shaped cells which infiltrated into fat tissue. There was no cellular atypia. All were compactible with desmoid-type fibromatosis of the omentum. Its was a sporadic case of omental fibromatosis with syndromal disease ruled out by subsequent investigations. At six months, there was no recurrence nor abdominal fibromatosis on clinical examination and in computer tomography image. No adverse effect was observed from surgical intervention.

**DISCUSSION:**

To the best of our knowledge, this is the second case of omental fibromatosis presenting as an irreducible inguinal hernia in the world, which the first case was also reported and published by our team in 1999 by Lam et. al [5]. In order to find out the incidence of the clinical presentation of this rare disease entity, we did a retrospective review with our hospital database. From 1.1.1996 to 30.6.2016, all hernia operations performed in all the affiliated Hospitals of the University of Hong Kong were reviewed retrospectively; data were retrieved from patient records inside clinical computer system.

A total of 7039 hernia operations were carried out during the period in which 564 were incarcerated or strangulated hernia operations, among which, 2 cases were of diagnosis of omental fibromatosis, which account for incidence of 0.028% of groin exploration of groin exploration. The incidence is extremely low according to our data, however there possibly patient with omental fibromatosis and yet operated in an earlier stage and therefore omental disease was not discover during time of operation. The actual incidence of omental fibromatosis with or without inguinal extension will be higher than what we have reported. In our patient, inguinal hernia was left untreated until it became symptomatic, leading to this unusual presentation as an incarcerated hernia.

Pathologically, fibromatoses is usually composed of spindle-shaped cells with keloid type collagen deposition, may stain positive for CD117 but is usually negative for CD34[2]. Atypical mitosis is basically absent and the amount of collagen is generally greater than that seen in GISTs. Fibromatosis often display cytoplasmic (not membranous) CD117 positivity. GISTs lack nuclear β-catenin while it is present in fibromatosis [6]. Wide surgical excision with clear margins is the treatment of choice. But there still is a tendency for local recurrence after radical surgery. Koh et. al. divided the clinical course into five categories: (1) spontaneous regression, (2) static, (3) variable growth, (4) progressive growth, and (5) aggressive growth [7]. Various treatment options can be considered includes radiotherapy, chemotherapy, antiestrogenic therapy, nonsteroidal anti-inflammatory drugs, and molecularly targeted therapy [2]. Omental fibromatosis or other form of inguinal mass or tumor should be

one of the differential diagnoses of an unusual presentation of incarcerated inguinal hernia.

## CONCLUSION:

Inguinal omental fibromatosis is rare and it could present as incarcerated hernia clinically. Fibromatosis may be part of presentation of syndromal disease like Garden's syndrome, the recurrence is higher than in sporadic cases despite radical surgery.

## Declaration of conflict of interest:

CT… declares no conflict of interest
JY… declares no conflict of interest
OSHL … declares no conflict of interest
KKN … declares no conflict of interest
ZHL… declares no conflict of interest
WLL… declares no conflict of interest
JKMF… declares no conflict of interest

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Figure 1: Computer Tomography Scan (Coronal Cut) showing the tumor mass extending to right scrotum

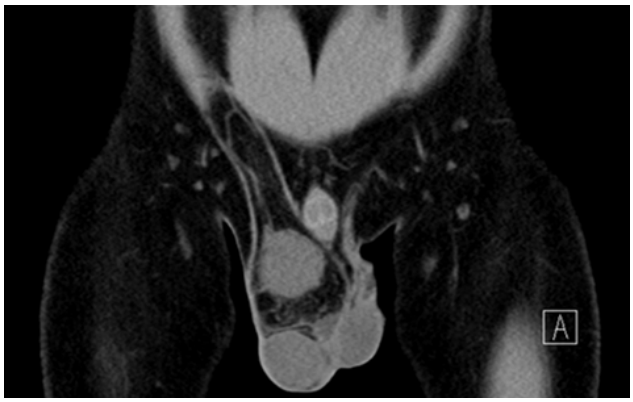



Figure 2: Computer Tomography Scan (Axial Cut) showing the tumor mass over the right inguinal canal

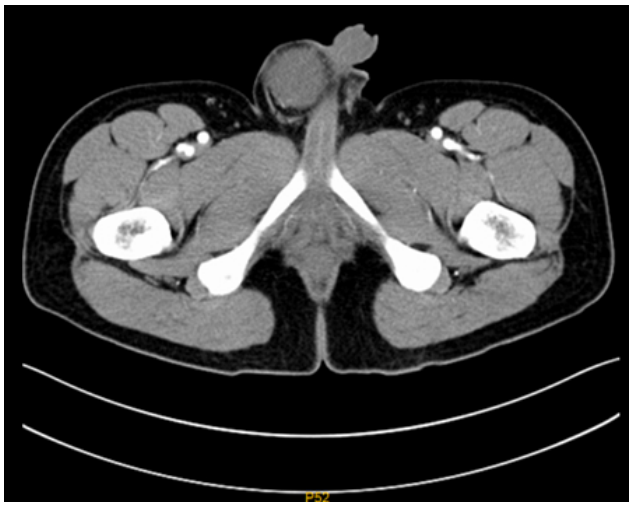


Figure 3: Excised omental fibromatosis forming a tumor mass

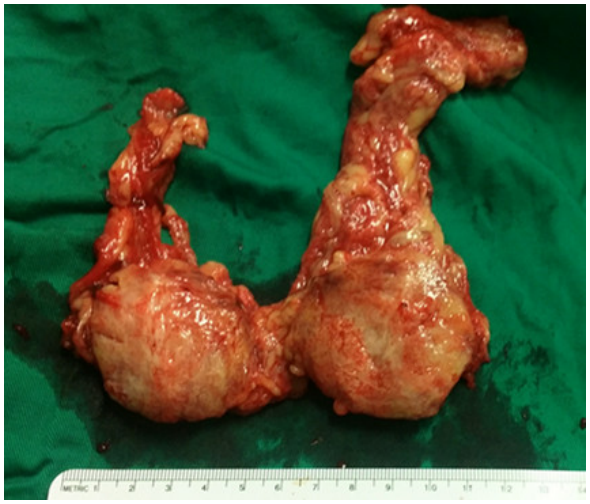
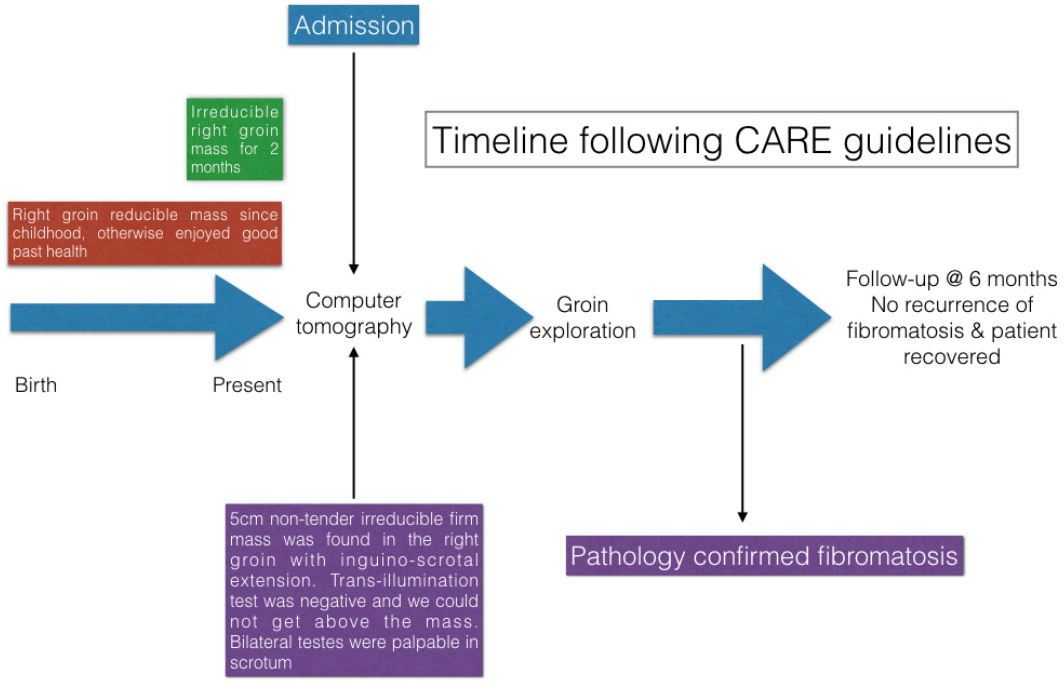


Figure 4: TIMELINE (following CARE guidelines)



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