EJCRIM
European Journal
of Case Reports
in Internal Medicine

A CASE REPORT OF MESENTERIC PANNICULITIS AND FIBROUS DYSPLASIA WITH PRIMARY METABOLIC SYNDROME

Birgit Khandalavala[1], Ryder Cuppett[2]

[1] Department of Family Medicine, University of Nebraska Medical Center, Omaha, USA
[2] College of Medicine, University of Nebraska Medical Center, Omaha, USA

Corresponding author's e-mail: birgit.khandalavala@unmc.edu

Received: 26/09/2024
Accepted: 06/11/2024
Published: 28/11/2024

Conflicts of Interests: The Authors declare that there are no competing interests.
Patient Consent: Written informed consent from patient and permission for publishing the patient's clinical history has been obtained. A signed patient consent has been included in the submission.
Acknowledgments: Patient personal involvement and consent to get her case published. Mary Beth Steele for manuscript preparation.

How to cite this article: Khandalavala B, Cuppett R. A case report of mesenteric panniculitis and fibrous dysplasia with primary metabolic syndrome. *EJCRIM* 2024;**11**:doi:10.12890/2024_004922

ABSTRACT

Introduction: Mesenteric panniculitis (MP) is a rare clinically diagnosed disease resulting from fibro-inflammation in the intestinal adipose tissue. Precedent intra-abdominal inflammation or trauma is often evident. Fibrous dysplasia (FD) is a benign uncommon childhood disease of the bones, with fibrous connective tissue replacing bone tissue. FD can cause pain, disfigurement, and entrapment of critical neighbouring structures. Adult-onset craniofacial involvement is rare. Metabolic syndrome is characterized by obesity and chronic subclinical inflammation, promoting abnormal cellular fibrosis and proliferation. The concomitant incidence of MP and FD in an older female adult with metabolic syndrome is unprecedented. An aberrant metabolic contributory pathophysiology for both MP and FD could be postulated.

Case Report: A 60-year-old Caucasian woman with primary long-standing well-treated metabolic syndrome with obesity presented for revision bariatric surgery. She had previous intra-abdominal surgeries and occasional intermittent abdominal pain. The surgery was abandoned due to extensively inflamed intestinal mesentery. Radiological studies and intra-abdominal biopsy confirmed MP. She subsequently developed pronounced unilateral facial swelling and radiological studies confirmed FD. Treatment has been suboptimal for both MP and FD. She developed venous thromboembolism on tamoxifen treatment for MP. The patient currently reports abdominal pain and swelling from MP, and facial disfigurement, pain, and tinnitus from FD. Blood markers for inflammation have remained consistently high, and there is statin intolerance. Most recently liver steatosis and diabetes have developed.

Conclusion: A case of MP and FD with primary metabolic syndrome is unique. An increased awareness of such rare cases could lead to further research, and critically needed treatments to optimize outcomes.

KEYWORDS

Mesenteric panniculitis, fibrous dysplasia, metabolic syndrome, obesity

EFIM
European Federation of Internal Medicine

Publisher
smc media srl

LEARNING POINTS

- Mesenteric panniculitis (MP) is a rare disease as is craniofacial fibrous dysplasia (FD) in adults. The incidence of these two rare diseases in an older adult with primary metabolic syndrome is previously unknown, making this case the first of its kind.
- Metabolic syndrome is common condition of the industrialized population, and an aberrant common "systemic" metabolic pathophysiology could contribute significantly to the inflammatory and fibrotic cellular changes inherent to both these two localized diseases of MP and FD.
- Knowledge about rare diseases, particularly if clustered with a primary systemic disease, could lead to better treatment outcomes in the future and promote novel therapeutic targets.

INTRODUCTION

Mesenteric panniculitis (MP) is a benign non-specific fibroinflammatory condition affecting the intestinal mesentery, rarely diagnosed clinically, and commonly presents with abdominal pain in the elderly[1,2]. MP can be asymptomatic and found incidentally[1]. Abdominal pain is the most frequent symptom present in 54-78% of patients with other symptoms much less common[1]. There is often precedent trauma or inflammation such as abdominal trauma, previous surgery, or autoimmune diseases, leading to the concept of MP as a "process of nonspecific inflammatory, autoimmune reactivity to any aggression or local inflammatory reaction in the abdomen"[2]. MP is diagnosed based on characteristic computed tomography (CT) imaging findings and histological studies[2]. There is no specific treatment for MP, and management is often guided by symptoms or empirical and individualized[1].

Fibrous dysplasia (FD) is another rare fibroproliferative disease, presenting most commonly in children, where atypical fibrous connective tissue replaces bone, skin, or connective tissue[3]. FD presents as polyostotic in early life and monostotic in later life. Post-natal skeletal stem cells proliferate resulting in fibro-osseous tissue that can cause fractures, deformity, pain, and functional impairment. The diagnosis of FD is made with radiological imagining and histopathology[3]. There is no definite treatment for FD, and modalities are conservative and palliative, with an emphasis on optimizing function and minimizing morbidity[3].

Metabolic syndrome is a cluster of comorbid conditions that indicate increased cardiovascular risk and traditionally includes hypertension, disordered carbohydrate, lipid metabolism, and atherogenic dyslipidaemia[4]. Metabolic syndrome is characterized by obesity and chronic subclinical inflammation[4]. Venous thromboembolism and fibrosis of the liver cells are considered part of the newly extended spectrum of metabolic syndrome. While individually MP and adult cases of craniofacial FD are rare, their concurrent occurrence in one patient, an older female patient with primary metabolic syndrome, is unprecedented, as evident in a review of the published literature (PubMed, Google Scholar, Medline, Cochrane, EMBASE). The aim of this unique case report is to increase awareness of these two rare diseases and to encourage further investigation into their aetiology, diagnosis, and treatment.

CASE DESCRIPTION

A 60-year-old Caucasian female with long–standing metabolic syndrome and a body mass index (BMI) of 43 kg/m^2 presented for elective revision bariatric surgery in 2017. Weight gain began in childhood, but continued throughout life, leading to two prior bariatric procedures in midlife (laparoscopic gastric band 2008 and laparoscopic gastric sleeve 2011). With increasing immobility due to chronic back pain and significant weight regain, revision bariatric surgery was elected. Blood pressure was well treated, but dyslipidaemia was persistent due to statin intolerance, and there was occasional intermittent abdominal pain with previous laparoscopic cholecystectomy. Surgery was abandoned on visualization of the extensive mesenteric adhesions to the abdominal wall, shortening of the mesentery, and interloop adhesions of the intestines. A biopsy of the abnormal mesenteric tissue was done, and histopathology revealed reactive epithelial changes of the mesentery and diffuse intraepithelial lymphocytes consistent with MP. An abdominal CT scan confirmed mildly prominent lymph nodes and characteristic central mesenteric fat infiltration consistent with MP *(Fig. 1)*. Family history was negative for MP or FD. A year later, the patient had worsening intermittent left-sided facial pain with ocular symptoms and progressive facial disfigurement, causing difficulty with wearing her glasses *(Fig. 2)*. A CT scan of the head in 2020 revealed a left orbital enlargement with possible multiple apparent osteochondromas along the left lateral orbit and squamosal temporal bone. Magnetic resonance imaging (MRI) in 2021 confirmed left FD of the zygoma with a "ground glass" internal appearance consistent with FD *(Fig. 3)*.

The differential diagnosis for chronic intermittent abdominal pain without bowel disturbances in a female adult in midlife with obesity includes, most often, malignancy, cholecystitis, bile duct cholecystitis, diverticulitis, and pancreatitis[1]. Previous cholecystectomy and radiological studies ruled these other conditions out. The fat masses could be lipomas, pseudo-tumours, or malignant tumours prior to definitive biopsy confirmation of MP. There is no specific treatment for MP and one of the initial treatments was tamoxifen, but this led to venous thromboembolism. Azathioprine was not tolerated and linaclotide was ineffective. Unilateral bony swelling of the face can be osteosarcoma, osteomyelitis,

and non-ossifying fibroma[3]. Radiological studies are confirmatory of FD and biopsies may not always be feasible. Currently, the patient is not undergoing active treatment for FD and is under surveillance.

Metabolic syndrome is a complex cardiometabolic condition, and the diagnosis is made based traditionally on central obesity, blood pressure, and blood metabolic markers[4]. The diagnosis has been recently broadened to include such diverse entities as chronic inflammation and raised inflammatory markers, sleep apnoea, tachycardia, liver, and heart failure with preserved-ejection fraction. Newer criteria using these expanded categories make it easy to diagnose metabolic syndrome in most adults, including most older adults living in industrialized countries.

The patient describes her current symptoms as abdominal pain and facial pain with trigeminal distribution, with a severity of 6 out of 10 at rest, with occasional abdominal swelling, chronic back pain with radiculopathy, constipation, depression, and poor quality of life. She is on treatment for sleep apnoea. New onset hearing loss is evident with possible auditory nerve involvement. An abdominal CT scan in 2021 showed persistent and unchanged MP with hepatic steatosis, and recently, diabetes type 2 has developed. With the recent availability of a new class of drugs, the glucagon-like peptide-1 (GLP-1) agonists, semaglutide followed by tirzepatide has been added. With this she has experienced some initial weight loss. The inflammatory marker, highly sensitive C-reactive protein continues to be elevated since 2017, between 16.1 to 29.7 mg/dl, with the latest 24.1 mg/dl in June 2024 (ideal level <3-1 mg/dl for average cardiovascular risk); the erythrocyte sedimentation rate has increased from 34 to 42 mm/hr (normal range 0-30 mm/hr) during this time range. There is no leucocytosis or anaemia. High-density lipoproteins (HDL) cholesterol has not been greater than 40 mg/dl with slightly increased triglycerides, and normal LDL cholesterol.

DISCUSSION

We describe the novel presentation of the concurrent appearance of two rare diseases, MP and craniofacial FD, in an older female adult, with primary progressive metabolic syndrome and persistently elevated inflammatory markers. We assume that the MP in this case was triggered by previous intra-abdominal surgeries, yet a systemic inflammatory cause cannot be excluded[5]. In keeping with recent findings that the incidence of MP is likely to be underestimated and that up to 50% of cases could be asymptomatic or found incidentally. Our case of extensive MP, found incidentally, demonstrates that underestimation is more likely the norm[1]. Our case also illustrates the frustrating course for most patients due to limited treatment options.

The FD that manifests in our case is atypical, since craniofacial FD is more common in children[3]. Considering the usual genetic aetiology of FD, the late and atypical presentation makes such a genetic causal relationship unclear and open to speculation. Similar to MP, it has also been noted that

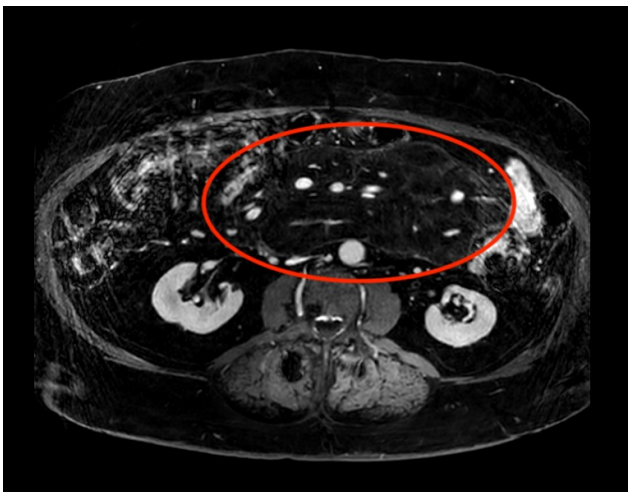

Figure 1. Axial contrast-enhanced computed tomography scan of the abdomen of a 60–year–old female with mesenteric panniculitis showing (circled area) the characteristic mass-like area of increased fat attenuation, with multiple prominent mesenteric lymph nodes and infiltration of the mesenteric fat centrally, displacing adjacent bowel segments.

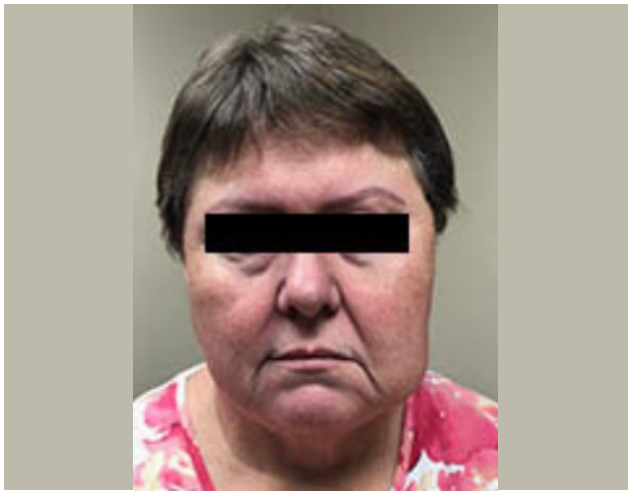

Figure 2. The 60- year-old female with prominent diffuse swelling of the left side of the face.

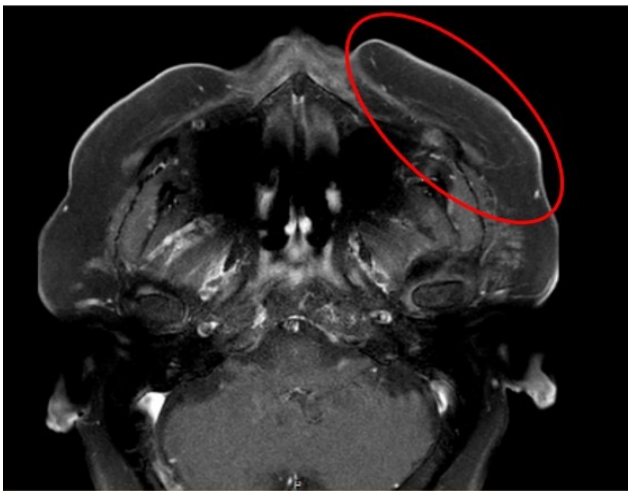

Figure 3. Magnetic resonance imaging of the head and orbits showing left zygoma involvement with characteristic findings of fibrous dysplasia of "ground glass appearance" with well-defined borders (circled area).

subclinical cases are likely to be much more common than previously known[3].

The cluster of conditions that constitute metabolic syndrome was prevalent for decades in our patient, probably beginning in childhood, and is progressive. Given that MP and FD are unlikely to be directly related but share common elements of localized cellular proliferation and inflammation. We postulate that the systematic influences of metabolic syndrome, particularly chronic inflammation, could be a significant and previously unrecognized contributory factor. One previous case of MP with primary Sjogren's has been described, however our patient did have a previous work up for rheumatological diseases that was negative[5]. Another similar case of FD in a patient with an unusual presentation in the pelvis with metabolic syndrome and subsequent diabetes has also been recently reported[6]. In our patient, decades of significantly raised inflammation from metabolic syndrome and obesity preceded the two rare diseases, making metabolic syndrome more than likely a significant and an unrecognized underlying factor. Metabolic syndrome has recently emerged as a significant association with MP and is noted to be present in 45% of patients with MP compared to 31.8% of controls in a study of 3698 patients with MP[1]. Metabolic syndrome is being seen increasingly in children and young adults and the comprehensive management of metabolic syndrome should begin much earlier in life than is currently being done, indicating a paradigm shift in the current standard practice of later treatment in adults. The true incidence of MP is likely to be much higher than reported since patients tend to be asymptomatic, and findings are seen incidentally on radiology testing. Since treatment options are limited, clinicians may not be that motivated to act on these findings of a usually benign condition[7]. Similarly, FD is often seen incidentally in radiological studies, but since there is no definitive treatment or patients are asymptomatic, no active management is started[3]. Our study indicates that such radiological findings should probably be given much greater clinical importance. Increasing the awareness of the medical team may be the critical first step in early diagnosis since advanced disease may be less amenable to successful treatment.

CONCLUSION

Our novel case study describes two rare diseases, MP and FD in a female adult with primary metabolic syndrome. Increased awareness, diagnosis, and research of both MP and FD appear warranted. Simultaneously, early and aggressive management of metabolic syndrome may be critical to optimizing long-term metabolic health and, eventually, quality of life and aging.

REFERENCES

1. Sulbaran M, Chen FK, Farraye FA, Hashash JG. A Clinical Review of Mesenteric Panniculitis. *Gastroenterol Hepatol (NY)* 2023;**19**:211-218.
2. Pinheiro F, Rgo A, ArajoFilho I. Mesenteric panniculitis in the elderly - update on diagnostic and therapeutic approach. *Int J Surg Med* 2016;**2**:127.
3. Kim HY, Shim JH, Heo CY. A rare skeletal disorder, fibrous dysplasia: a review of its pathogenesis and therapeutic prospects. *Int J Mol Sci* 2023;**24**:15591.
4. Dobrowolski P, Prejbisz A, Kuryłowicz A, Baska A, Burchardt P, Chlebus K, et al. Metabolic syndrome—A new definition and management guidelines. *Arter Hypertens* 2022;**26**:99–121.
5. Mendes PA, Ferreira DM, Temido H, Pina R, de Carvalho A. Mesenteric panniculitis in Sjögren's syndrome: a new systemic manifestation to consider? *Eur J Case Rep Intern Med* 2018;**5**(4).
6. Alkhaibary A, Alassiri AH, Alsalman M, Edrees S. Unusual presentation of fibrous dysplasia in an elderly patient. J Radiol Case Rep 2019;13:26.
7. Eze VN, Halligan S. Mesenteric panniculitis: a clinical conundrum. *Br J Radiol* 2023;**96**:20211369.