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RADIOLOGY CASE REPORTS

Case Report

Transarterial doxorubicin-eluting beads embolization for the treatment of desmoid fibromatosis in the pubic region ☆

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ABSTRACT

Desmoid fibromatosis (DFs) is rare, low-grade neoplasm. Although it poses no risk of metastasis, DFs exhibits a range of clinical manifestations characterized by local infiltrative growth tendencies and a propensity for recurrence. Despite its nonmalignant nature, DFs can be highly debilitating and occasionally life-threatening, causing severe pain and functional limitations. Traditionally, surgery served as the conventional primary treatment approach; nevertheless, a recent shift in paradigm towards a more conservative management has emerged, accompanied by efforts to standardize the strategy among clinicians. Systemic doxorubicin has been demonstrated to be effective in treating DFs; however, it carries potential risks of adverse effects on the cardiovascular, digestive, and hematologic systems. The novel intravascular intervention employing drug-eluting beads loaded with doxorubicin represents an effective treatment for DFs, optimizing drug delivery to the target lesion and reducing systemic toxicity. In this article, we present a rare case of DFs in the right pubic region treated with transarterial doxorubicin-eluting beads embolization.

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Introduction

According to the World Health Organisation (WHO), DFs is a "clonal fibroblastic proliferation that arises in the deep soft tissues and is characterised by infiltrative growth and a tendency toward local recurrence but an inability to metastasise" [1]. DFs is rare, with an annual incidence rate of 5-6 cases per 1 million individuals, constituting <3% of all soft-tissue tumors and approximately 0.11% of all bone tumors, with a peak age of 30-40 years [2–7]. Due to its infrequency, as many as 30%-40% of cases may be misdiagnosed, leading to inappropriate or delayed medical attention [4,8]. DFs can be categorized into 2 groups: those associated with familial

adenomatous polyposis (FAP) or gardener's syndrome, and those that occur sporadically in individuals without FAP, with the latter representing about 85% of cases and exhibiting a female predominance [9]. DFs can impact any region of the body, but they are more commonly located in the extremities for sporadic cases and intra-abdominally for individuals with FAP. Accumulating evidence demonstrating long-term stabilization or spontaneous regression in numerous patients with sporadic DFs has prompted a shift in the paradigm from immediate surgical resection to more conservative approaches, notably active surveillance [10,11]. Intervention is warranted in cases of progressive symptoms or sustained interval growth [2,4]. Systemic therapies encompass a diverse array, spanning from the use of nonsteroidal anti-inflammatory drugs and hormonal treatments to targeted tyrosine kinase inhibitors and cytotoxic chemotherapy [12]. Multicenter retrospective reviews have revealed that DFs treatment regimens involving doxorubicin are associated with better outcomes [13,14]. Despite its promise, systemic doxorubicin carries side effects, including palmar-plantar erythema, mucositis, and dose-related cardiotoxicity [15,16]. Selective transarterial embolization with doxorubicin-eluting beads was employed to optimize local efficacy and reduce systemic toxicity in treating DFs [17]. This article showcases a rare case of desmoid fibromatosis in the right pubic region, treated through transarterial embolization with doxorubicin-eluting beads.

Case report

A 50-year-old female patient admitted to hospital due to pain in the right pubic region. Six months before admission, she fortuitously detected a mass in the right pubic area, which underwent biopsy. The pathological examination result was desmoid fibromatosis. Throughout the course of the illness, the patient experienced a lingering pain in the right pubic region, which radiated down to the inner surface of the right thigh, with the intensity of the pain gradually increasing over time. Upon admission, the patient exhibited severe pain that substantially hindered mobility, daily activities, and work, registering a score of 6 points on the Visual Analogue Scale (VAS). Laboratory tests, including complete blood count, coagulation profile, electrolytes, urinalysis, blood glucose, liver function, and renal function, were within normal limits.

The pelvic radiograph revealed osteolytic lesions in the right pubic region, affecting both the superior and inferior pubic ramus, characterized by an indistinct, irregular margin without periosteal reaction visualized (Fig. 1). On the pelvic computed tomography (CT) scan images, lytic bone lesions were observed at the superior and inferior pubic ramus and the anterior surface of the right acetabulum, resulting in cortical disruptions and extension into the adjacent soft tissue (Fig. 2). On the MRI, these lytic bone lesions were hypointense on T1-weighted images, hyperintense with some areas of hypointense on T2-weighted images, and showed intermediate to high signal intensity on short tau inversion recovery (STIR) images, with heterogeneous contrast enhancement (Fig. 3).
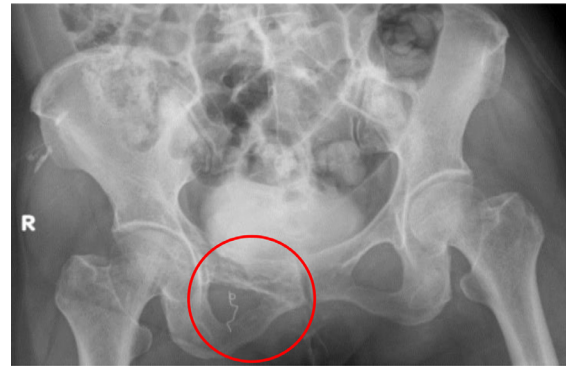

Fig. 1 – On the pelvic radiograph, osteolytic lesions, affecting both the superior and inferior pubic ramus (circle), characterized by an indistinct, irregular margin without periosteal reaction visualized.

The patient underwent transarterial doxorubicin-eluting beads embolization for the treatment of DFs. She received localized anesthesia, following which a 5F Sheath was introduced into the left femoral artery using the Seldinger technique. Subsequently, sequentially insert a Pigtail catheter 5F and a Cobra catheter 5F (Merit Medical, USA) into the right common iliac artery and internal, external iliac arteries, acquiring digital subtraction angiography (DSA) diagnostic images, demonstrating a vascular proliferation mass in the right pubic region, receiving blood supply from pubic branches of the right obturator artery.

Selective embolization was conducted using a 2.6 F microcatheter passed over a 0.021-inch guide wire (Asahi Masters Parkway HF kit, Asahi Intecc, Japan). Select the posterior branch of the right obturator artery and deploy 01 coil to mitigate blood flow into this branch. Subsequently, selective embolization the pubic branches from the anterior branch of the right obturator artery, using doxorubicin one vial of Hepasphere 30-60 μm (Merit Medical, USA), which was loaded with 75 mg doxorubicin (Fig. 4). Post embolization DSA images confirmed successful occlusion of the proliferating pubic branches originating from the right anterior obturator artery, the procedure was then concluded.

After the interventional procedure, the patient manifested a diminution in discomfort within the right pubic region, enhanced mobility, and increased ease in participating in routine daily activities as compared to the preinterventional state. The pain level at the time of discharge was approximately 2 points based on the Visual Analog Scale (VAS). No complications were observed postprocedure. On the MRI images obtained for postinterventional evaluation at 8 months, the volumetric reduction of the lesion was observed to be approximately 58.1%. The maximal diameter exhibited a reduction of 10.2%, accompanied by decreased signal intensity on T2-weighted imaging and reduced contrast enhancement compared to preinterventional assessments (Fig. 5). Furthermore, there was an absence of soft tissue component postintervention. The lesion was categorized as stable disease (SD) in accordance with revised Response Evaluation Criteria in Solid
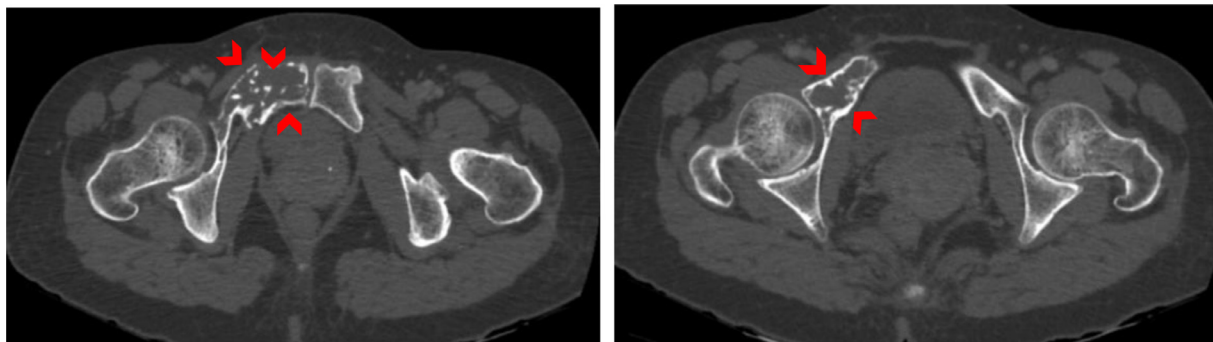
Fig. 2 – On the CT scan images, lytic bone lesions were observed at the superior and inferior pubic ramus and the anterior surface of the right acetabulum, resulting in cortical disruptions and extension into the adjacent soft tissue (arrowheads).
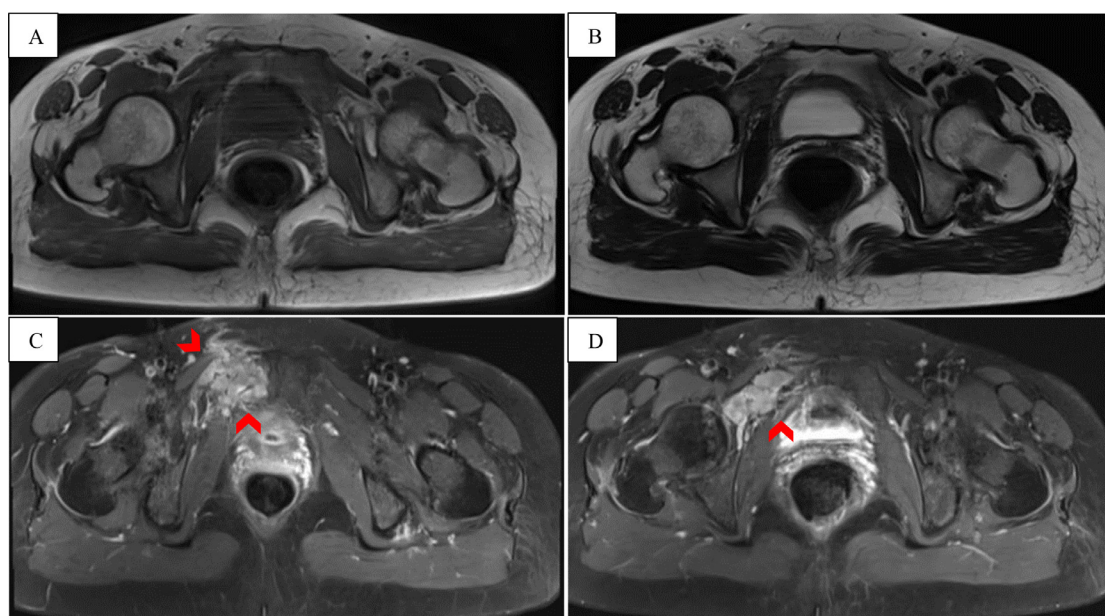

Fig. 3 – On the MRI, these lytic bone lesions were hypointense on T1-weighted images (A), hyperintense with some areas of hypointense on T2-weighted images (B), and showed heterogeneous contrast enhancement (arrowheads, C and D).

Tumors (RECIST 1.1) criteria [18]. Clinically, the patient's condition remained stable, with a pain score of approximately 2 points based on the Visual Analog Scale (VAS), allowing normal daily activities and work.

Discussion

DFs is uncommon, manifesting with an annual incidence rate of 5-6 cases per 1 million individuals, comprising less than 3% of all soft-tissue tumors and approximately 0.11% of all bone tumors [2,4–7]. Despite exhibiting some variability, there is a notable 2- to 3.5-fold increased incidence observed in women [12,19,20]. A diverse age spectrum is impacted, with the majority of instances occurring within the age range of 15 to 60 years, and an average age of 36.7 years [12,20]. DFs can be intra-abdominal, within the abdominal wall, or extra-abdominal. Across various studies, 28%-69% of desmoids were intra-abdominal or abdominal wall, with the remaining cases were characterized as extra-abdominal [21]. Extra-abdominal DFs are closely linked to muscle and fascial tissues, predominantly occurring in the shoulder and upper extremity (33%), gluteal region and lower extremity (30%), as well as the chest wall or back, and the head and neck (10%) [6,22–24]. Compare to soft tissue DFs, desmoplastic fibroma of the bone is even rarer. Due to its infiltrative growth pattern, bone DFs has been regarded as the osseous counterpart of extra-abdominal DFs, owing to its resemblance in histological characteristics [25]. Therefore, the pubic bone desmoid fibromatosis as described in this article represents an exceptionally rare case.

A wide spectrum of treatments is available for DFs, encompassing local therapies such as surgery, radiotherapy (RT), percutaneous treatments, and an expanding list of efficacious systemic therapies. This diversity of therapeutic options enables substantial customization of treatment regimens
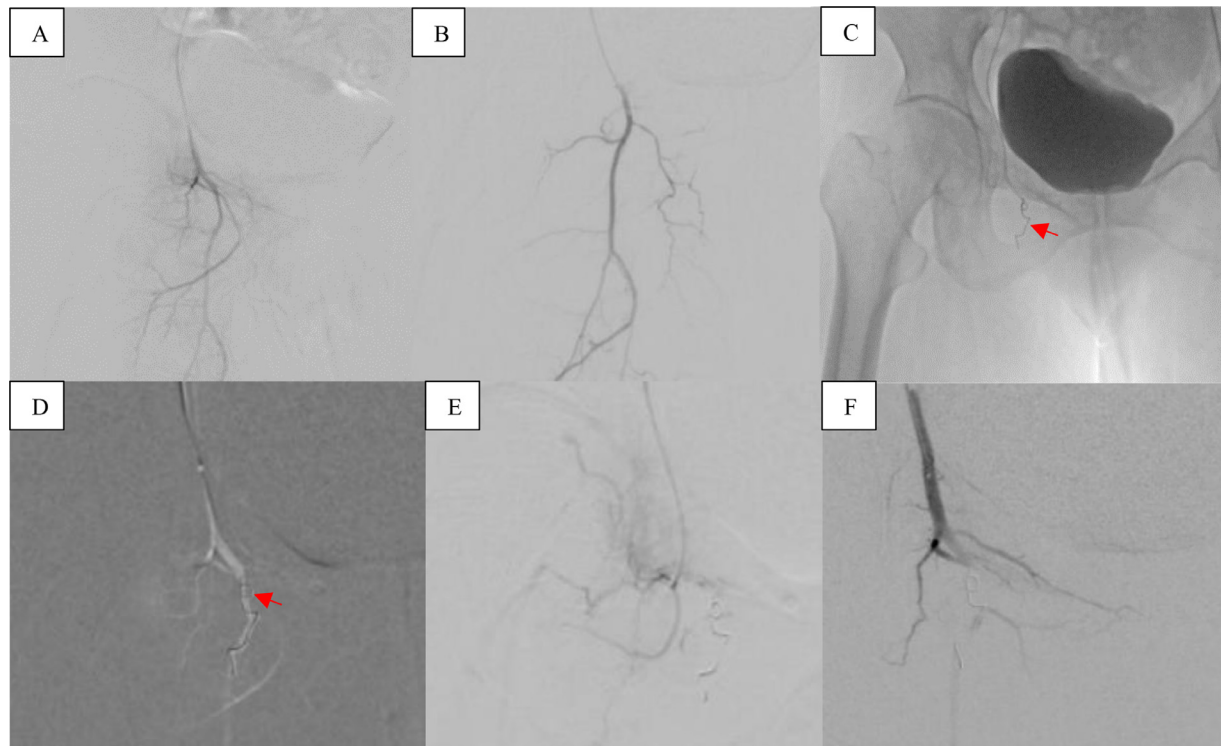
Fig. 4 – Selective embolization was conducted using a 2.6 F microcatheter (A). Select the posterior branch of the right obturator artery (B) and deploy 01 coil to mitigate blood flow into this branch (C and D, arrow). Subsequently, selective embolization the pubic branches from the anterior branch of the right obturator artery, using doxorubicin drug-eluting beads (D and E). Post embolization DSA images confirmed successful occlusion of the proliferating pubic branches originating from the right anterior obturator artery (F).

tailored to the individual patient. Historically, surgical wide-margin resection has been considered the primary approach for managing DFs. Nevertheless, rates of postsurgical local recurrence at 5-10 years have been reported to fall within the range of 30% to 77% [26–28]. Microscopic margin-negative (R0) resections were not achieved in most surgeries, and there was no consensus on whether a positive margin resection correlated with the risk of recurrence [4,29]. The rates of postsurgical relapse were found to be higher in cases of extra-abdominal DFs compared to those in abdominal DFs [30]. Radiation therapy has been employed in the management of DFs either as an adjunct to surgery or as the primary treatment modality in cases where tumors are unresectable or where resection would substantially compromise functional outcomes [31,32]. Given that patients diagnosed with DFs are frequently younger and exhibit a favorable long-term survival prognosis, it is imperative to weigh the potential complications associated with radiation therapy. These complications may include limb contractures, pain, and the development of in-field skin cancer [33,34].

Several retrospective series provide evidence endorsing the utilization of systematic doxorubicin and doxorubicin-based combination in the management of patients diagnosed with DFs [16,21,35]. Nevertheless, the significant toxicities linked with doxorubicin-based treatment regimens, particularly cardiotoxicity and myelosuppression, represent major concerns [36]. According to recent studies, transarterial

doxorubicin-eluting beads embolization is a novel treatment method that optimizes treatment effectiveness locally with significantly lower doses of doxorubicin compared to systemic dosing, which may help mitigate the adverse effects [17,37,38]. A combined prospective and retrospective study found that doxorubicin-eluting beads embolization was effective, resulting in a significant reduction in lesion signal intensity on T2-weighted images and a median volume decrease of 59% after 8 months follow-up; additionally, 52% of patients achieved stable disease according to RECIST 1.1 criteria [17]. The relationship between the T2 signal intensity and the tumor's hypo-intense areas is indicative of the relative proportions of cellular density to fibrotic matrix within DFs [39]. The evaluation of DFs is not optimally performed through 2-dimensional measurements. Thus, alterations in volumetric measurements offer a more precise means of assessing response compared to merely assessing the lesion's largest dimension. Hence, reductions in tumor volume and T2 signal intensity serve as dependable indicators of treatment efficacy. Transarterial doxorubicin-eluting beads embolization is also safe, and well-tolerated, with the mean doxorubicin dose was significantly lower than the typical systemic dose [17,37,38]. In our case of pubic bone DFs, we conducted coiling embolization to diminish blood flow to the posterior branch of the right obturator artery, followed by superselective doxorubicin-eluting beads embolization of the pubic branches originating from the anterior branch of the

Fig. 5 – MRI images obtained for postinterventional evaluation at 8 months. The lesion's maximal diameter exhibited a reduction of 10.2% (E: preprocedure, F: 8 months follow-up), accompanied by decreased signal intensity on T2-weighted imaging (circle, A: preprocedure, B: 8 months follow-up) and reduced contrast enhancement compared to preinterventional assessments (C and D). Furthermore, there was an absence of soft tissue component postintervention (arrows).

right obturator artery. The objective was to optimize targeted tumor treatment while minimizing nontarget embolization.

Conclusion

In this article, we present a particularly rare case of desmoid fibromatosis in the right pubic region, which responded to transarterial doxorubicin-eluting beads embolization. Our case demonstrates the safety and efficacy of this promising treatment approach, especially for cases of inoperable extra-abdominal desmoid fibromatosis or those unsuitable for other local therapies.

Patient consent

Informed consent for patient information to be published in this article was obtained.

Ethical statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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