Case Report

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Ectopic primary ductal breast carcinoma of the vulva: a case report and literature review

Marah Mansour, MD[a,b], Orgwan Zahra, MD[g], Dalia Nabulsi, MD[h], Aya Alhamwi, MD[c], Maya Chahin, MD[c], Wael Raghid alani, MD[b], Diaa Yousef, MD[e], Ilda Kanbour, MD[d], Bashar Kurdi, PHD[d], Ahmad Mohammad Deeb, MS[f,*]

Background: Ectopic breast tissue (EBT) is normally found along the rudimentary mammary line, which is located from the axilla to the inguinal region and disappears during embryogenesis. Up to 6% of females around the world suffer from EBT. EBT cancer is rare, with only a few reports in the literature. The diagnosis, and treatment of these rare cases can be very difficult and complex.
Case presentation: The authors report an interesting case of a 74-year-old female patient with a vulvar lump located on the mons pubis associated with ulceration, pain, and size increase. The histopathological study demonstrated an invasive ductal carcinoma grade 2 arising from EBT in the vulva. Within 4 months of follow-up, the patient's condition was stable.
Clinical discussion and conclusions: Throughout this paper, the authors aim to highlight the diagnostic and therapeutic challenges since there are no current guidelines for treatment for such cases. Furthermore, this article describes this rare disease and includes surgical details, difficulties, diagnostic methods, and treatment options.

Keywords: case report, ductal carcinoma, Ectopic breast tissue, review article, vulvar lump

Background

Ectopic breast tissue (EBT) can appear anywhere along the milk line, which runs bilaterally from the axilla to the groyne. EBT develops as the result of inadequate regression of the mammary ridge during embryologic development. Females often have 1–6% EBT, which is more prevalent in the axilla or on the thorax but uncommon in the vulva. According to legend, Hartung reported the first instance of a completely developed mammary gland in a 30-year-old female's left labium majus in 1872; however, infiltrating ductal cancer resulting from EBT in the vulva was originally documented by Greene and colleagues in 1936[1–3]. EBT can exhibit benign or malignant breast pathology because it reacts to physiological stressors and hormonal stimuli similarly to breast tissue that is ordinarily found in the breast[4,5]. The typical clinical manifestation of EBT carcinoma is a fixed, palpable lump with irregular borders that, in 45% of instances, appears as a subcutaneous lesion in the labia majora[5,6]. Other symptoms are discoloration, ulceration, or bleeding from this mass, and in rare cases, an abscess. According to studies, the average age at diagnosis was 51 years[6]. As is the case with our patient, ductal carcinoma is the most common histological form in 79% of cases of ectopic breast cancer (EBC), followed by lobular, medullary, and other subtypes[6,7]. Investigations include both mammography and ultrasound, but a definitive diagnosis is made by a biopsy of the lesion[7]. Initially, the surgical procedure involved a mastectomy, the removal of the EBT, and a sentinel lymphadenectomy, while in advanced cases, radiotherapy and chemotherapy should be performed[7,8]. Throughout this paper, in light of a literature review, we describe a case of a 74-year-old female who had primary breast cancer in vulval supernumerary tissue at the time of presentation. This case has been reported in line with the SCARE criteria[9].

HIGHLIGHTS

- Ectopic breast cancer of the vulva is a rare disease.
- It is crucial to differentiate primary breast cancer of the vulva from metastatic cancer in this area.
- To establish the diagnosis, thorough history, physical examination, and diagnostic tests should be done.
- Diagnosis and management guidelines should be developed for primary ductal adenocarcinoma of the mammary-like glands of the vulva.

Case presentation

A 74-year-old female patient was admitted to the Department of Obstetrics and Gynecology with a complaint of a 4-year history of a vulvar lump, located on the mons pubis, that gradually increased in size. The lesion was ulcerated, painful, measured

[a]Department of Colorectal Surgery, Mayo Clinic, Rochester, MN, [b]Faculty of Medicine, Tartous University, Tartous, [c]Faculty of Medicine, Damascus University, [d]Department of Gynecology, Obstetrics University Hospital, Damascus, [e]Faculty of Medicine, Aleppo University, Aleppo, [f]Faculty of Medicine, Tishreen University, Latakia, Syrian Arab Republic, [g]Faculty of Medicine, Ankara University, Ankara, Turkey and [h]Faculty of Medicine, Kuwait University, Jabriya, Kuwait

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*Corresponding author. Address: Faculty of Medicine, Tishreen University, Latakia, Syrian Arab Republic. Tel.: +963945618531. E-mail: ahmadkhdeeb@gmail.com (A. Mohammad Deeb).

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15 cm, and pink in colour (Fig. 1). The patient's past medical history included hypertension for 12 years. No family history, surgery, or allergies. The physical examination was unremarkable, with no lymph node enlargement. Ultrasound findings were within normal limits (Fig. 2). The chest-abdomen-pelvic computed tomography (CT) scan showed a cystic lesion within the umbilicus measuring 2 × 3 cm with a hypodense lesion at the level of the fourth segment of the liver, the collapse of the body of the ninth dorsal vertebra with slight retraction of its posterior edges, and degeneration of the edges of the pubic symphysis with right-sided sclerosis without any lymphadenopathy or metastasis. Under regional anaesthesia, an excisional biopsy was performed (Fig. 3). The lump was cut with cautery and sewed by Vicryl filament 2-0 without using a slide, and the bleeding has been stopped. Histopathological studies confirmed the diagnosis of invasive ductal carcinoma grade II. Margins were negative for cancer. The diagnosis of a primary EBT of the vulva was established. The patient made a full recovery at the site of the operation, with follow-up at a cancer centre. The patient had no special concerns about the surgical procedure.

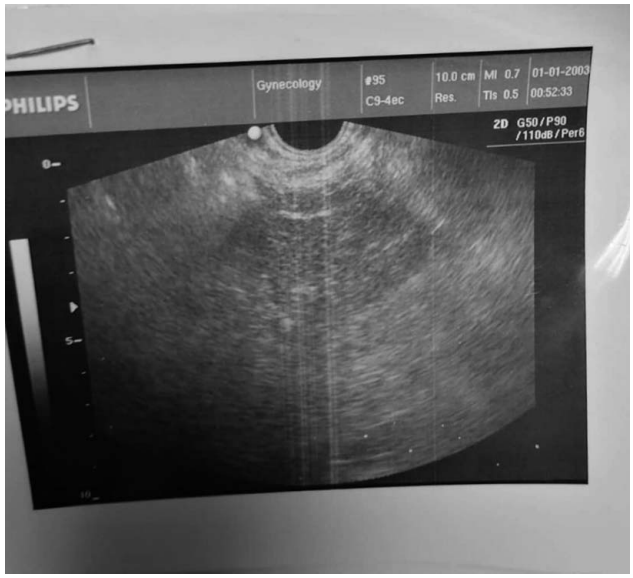

Figure 2. Ultrasound showing normal findings.

Discussion

Vulvar EBC is a rare disease. In general, EBT can be found at any site along the primitive milk line that extends from the axilla to the groyne. Unlike the vulva, the axilla represents the most common site of EBT. EBT can undergo different benign or malignant pathological changes. Thus, EBC may occur[1]. EBC represents 0.2–0.6% of all breast cancers[6]. Any histological subtype of breast cancer may occur in the vulva. Reports show that the ductal type was the most frequent subtype (42%)[3]. EBC patients present at ages between 45 and 82 years[5].

The aetiology of EBT in the vulva is still controversial[3]. However, there are 3 suggestive theories for its origin. The first is that EBT results from a failure of the embryonic mammary ridges to regress during embryogenesis. The second one suggests that human mammary ridges never reach the vulva during embryological development and that the anogenital mammary glands originate from anogenital tissue. The third one is that it may arise from pluripotent cells within the anogenital area[8].

Most EBCs of the vulva begin as asymptomatic nodules for an extended period before growing in size. There could be no other symptoms associated with the nodule, but some cases present with pain[1,8,10], discomfort[3,11–15], discharge[16], bleeding[1,11,16–18], dysuria[12], and swelling[1,19,20]. Initially, the diagnosis focuses on detecting a metastasis through a thorough history, physical examination, and a metastatic work-up that includes

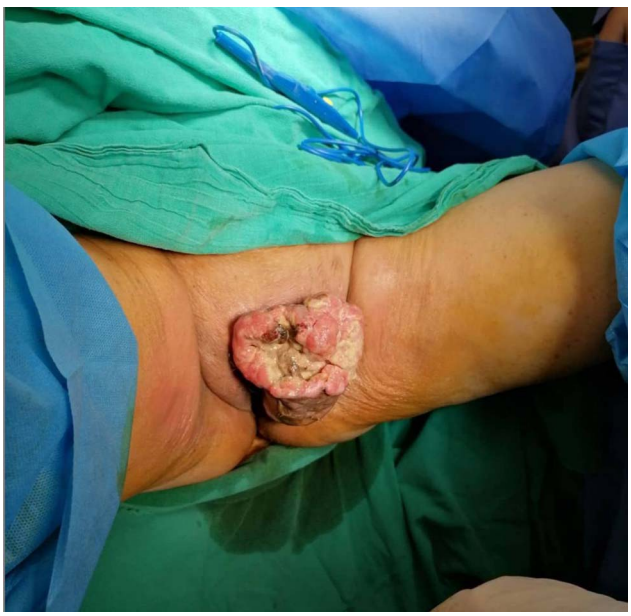

Figure 1. Vulvar lump located on mons pubis, ulcerated and pink in colour.

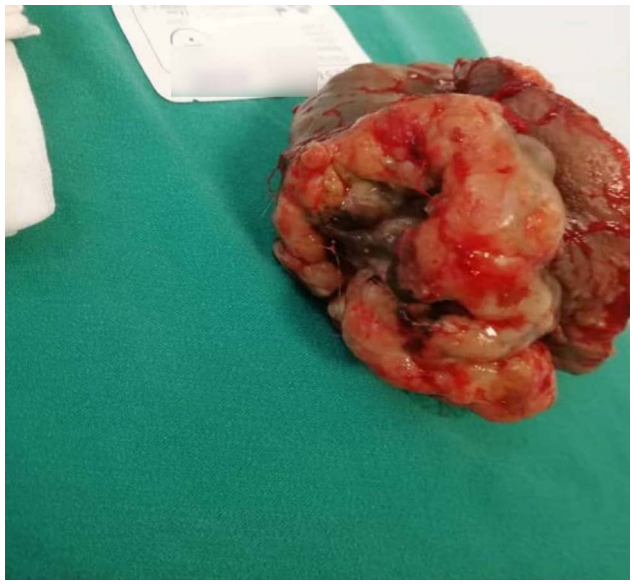

Figure 3. Benign glandular-like-mammary tissue (primary ectopic breast cancer of the vulva).

Table 1

Characteristics and management of primary breast ductal adenocarcinoma of the vulva

Ref	First author/PY	Age (year)	Clinical manifestations	Diagnosis	Receptors	Management	Outcome (month)
1	Greene *et al.* 1936[16]	49	(20 × 15) cm mass on R labia majora for 1 year, Bleeding, discharge, ulceration	Adenocarcinoma	N/A	N/A	DOD
2	Hendrix Behrman *et al.* 1956[10]	58	Painful 3 cm nodule on L labia minora for 5 years, Oedema, infection	Adenocarcinoma	N/A	Lymph nodes Biopsy: N/A Surgery: radical vulvectomy. Adjuvant therapy: N/A	N/A
[a]3	Gurrey Prat *et al.* 1976[35]	62	1.5 cm nodule, L labia major	DCIS, invasive ductal carcinoma	N/A	Lymph nodes biopsy: N/A Surgery: WLE Adjuvant therapy: N/A	DOD (24)
4	Cho *et al.* 1985[29]	70	(3 × 4 × 4) cm mass on R labia majora for 2.5 years	Infiltrating ductal carcinoma	ER +, PR +	Lymph nodes Biopsy: 2/9 Surgery: Radical R hemivulvectomy, ipsilateral LND. Adjuvant therapy: Hormonal therapy (Tamoxifen)	Alive (24)
5	Simon *et al.* 1988[11]	60	(2 × 2) cm lesion on R labia majora for 4 years, bleeding, discomfort	Adenocarcinoma	ER +, PR +	Lymph Nodes biopsy: 3/11 Surgery: Radical vulvectomy, bilateral LND. Adjuvant therapy: RT, Chemo, Tamoxifen and hormonal therapy on discharge	DOD(27) metastatic
6	Rose *et al.* 1990[22]	68	(2 × 2) cm mass, on R labia majora for 3 years	Infiltrating ductal carcinoma	ER + / PR +	Lymph Nodes biopsy: 1/15 Surgery: Radical vulvectomy, ipsilateral LND Adjuvant therapy: RT, Hormonal therapy (Tamoxifen)	N/A
7	Pelosi *et al.* 1991[32]	40	Tender 1.5 cm nodule on L labia majora for 3 months	DCIS	ER +, PR +	Lymph Nodes biopsy: N/A Surgery: Local Excision Adjuvant therapy: None	Alive (24)
8	Di Bonito *et al.* 1992[23]	46	1.5 cm nodule and 0.8 cm in thickness on R labia majora for 2 years	Infiltrating ductal carcinoma	N/A	Lymph Nodes biopsy: 11/13 Surgery: Radical vulvectomy, bilateral LND Adjuvant therapy: N/A	Alive (4)
9	Agha RA *et al.* 2020[9]	N/A	N/A	N/A	N/A	N/A	N/A
10	Bailey *et al.* 1993[17]	65	(3 × 2 × 1) cm nodule, R labia majora and labia minora for 3 years, Ulceration, bleeding	Infiltrating ductal carcinoma	ER +, PR +, GCDFP-15 +	Lymph nodes biopsy: R 2/10 Surgery: Radical vulvectomy, bilateral LND. Adjuvant therapy: Tamoxifen	Alive (12)
11	Van der Putte and van Gorp *et al.* 1994[33]	51	(1 × 1.5) cm nodule between R labia majora and minora for 3 months, erosion	Adenocarcinoma	CEA +, CAM5.2 +	Lymph Nodes biopsy: N/A Surgery: Excision and re-excision Adjuvant therapy: N/A	N/A
12	Levin *et al.* 1995[25]	62	2.5 cm nodule in L periclitoral region	Infiltrating ductal carcinoma	ER +, PR − HER2/NEU +	Lymph Nodes biopsy: 4/11 Surgery: WLE, ipsilateral LND. RT: NA Adjuvant therapy: Tamoxifen	N/A
13	Kennedy *et al.*, 1997[12]	71	(5 × 2) cm mass on L labia major for 1 month, lower abd discomfort, burning on urination	Infiltrating ductal carcinoma	ER −, PR −	Lymph Nodes biopsy: 9/9 Surgery: Radical vulvectomy, bilateral LND Adjuvant therapy: RT, Chemo	Alive (15)
14	Irvin *et al.* 1999[19]	64	Swelling, indurated (2.7 × 1) cm nodule on L mons pubis for 4 years	Invasive ductal carcinoma, invasive lobular carcinoma	ER +, PR +	Lymph Nodes biopsy: 1/14 Surgery: WLE, ipsilateral LND Adjuvant therapy: Chemo, RT, Hormonal Therapy (Tamoxifen)	Alive (36)

15	Gorisek *et al.* 2000[30]	81	Ulcerated (2 × 3) cm mass on L labia majora	Ductal	ER +, PR +	Lymph Nodes biopsy: NA Surgery: WLE Adjuvant therapy: Hormonal Therapy (Tamoxifen)	Alive (19)
16	Piura *et al.* 2002[24]	69	Ulcerated (3 × 3 × 3) cm nodule on L labia majora	Ductal	ER +, PR +	Lymph Nodes biopsy: 7/15 Surgery: Radical vulvectomy, bilateral inguinal LND Adjuvant therapy: Chemo, RT, Hormonal therapy	Alive (14)
17	Castro and Deavers *et al.*, 2001[34]	57	Ulcerated 2.5 × 0.7 cm nodule on R labia majora for 1 year	DCIS	ER +, PR +,	Lymph Nodes biopsy: SLN negative Surgery: WLE, re-excision Adjuvant therapy: letrozole, AI	Alive (12)
18	Ohira *et al.* 2004[15]	82	(5 × 3) cm mass on L labia major for 10 years, discomfort	Ductal	ER +	Lymph Nodes biopsy: infiltrated Surgery: WLE, L ipsilateral LND Adjuvant therapy: N/A	Alive (20)
19	Tanaka *et al.* 2005[13]	87	4 cm nodule on L labia majora for 3 years, discomfort	Adenocarcinoma	ER –, PR –	Lymph Nodes biopsy: N/A Surgery: No Surgery performed Adjuvant treatment: chemo: paclitaxel for 20 weeks, the patient refused further treatment	Mets in costa, sternum, vertebrae, Refused F-U
20	Intra *et al.* 2006[31]	53	Tender, reddish, 2 cm nodule on L abia major for 3 years	Ductal	ER +, PR +, HER2 –	Lymph Nodes biopsy: SLN 0/2 Surgery: WLE Adjuvant therapy: Hormonal Therapy (Tamoxifen)	Alive(20)
21	Abbot *et al.* 2006[20]	51	Tender 1 cm nodule on R labia majora for > 1 year, Swelling	Infiltrating ductal adenocarcinoma	ER +, PR +, HER2 –	Lymph Nodes biopsy: N/A Surgery: WLE, Mohs micrographic surgery. Adjuvant therapy: N/A	Alive (26)
22	Martines Palones *et al.* 2007[26]	49	(3.5 × 3) cm mass on R labia majora	Ductal	ER +, PR +	Lymph Nodes biopsy: 0/14 Surgery: Ipsilateral LND, WLE Adjuvant therapy: Hormonal therapy (Tamoxifen)	N/A
23	Tseung *et al.* 2008[14]	49	Nodule on R labia majora near clitoral hoos for 2–3 months, Discomfort	Infiltrating Ductal carcinoma	ER +, PR +	Lymph Nodes biopsy: Infiltrated Surgery: N/A Adjuvant therapy: N/A	N/A
24	Naseer *et al.* 2011[27]	57	painless 1.5 cm nodule on R labia majora for 2–3 years	Invasive Ductal carcinoma	ER +, PR +, HER2 –	Lymph Nodes biopsy: 3/13 Surgery: WLE, bilateral LND Adjuvant therapy: RT, Chemo, AI	Alive
25	Bogani *et al.* 2013[8]	71	Painful, ulcerated 4 cm nodule on L labia majora	Ductal carcinoma	ER +, PR +	Lymph Nodes biopsy: 1/ 8 Surgery: Radical vulvectomy, LND Adjuvant therapy: Chemo: 5 cycles, Hormonal therapy (Tamoxifen)	Alive (24)
26	Benito *et al.* 2013[21]	82	Ulcerated 4 cm nodule on L labia for 1 year	Invasive Ductal carcinoma	ER +, PR +, HER2 –	Lymph Nodes biopsy: 27/32 Surgery: Radical vulvectomy, LND Adjuvant therapy: AI	Alive (24)
27	Cripe *et al.* 2015[28]	62	1.3 cm nodule on L side of vulva for 1 year	Invasive ductal carcinoma	ER +, PR +, HER2 –	Lymph Nodes biopsy: 0/14 Surgery: WLE, LND Adjuvant therapy: N/A	Recurrence. WLE, LND, 3 lymph nodes + followed by CT, RT, and AI

Table 1
(Continued)

Ref	First author/PY	Age (year)	Clinical manifestations	Diagnosis	Receptors	Management	Outcome (month)
28	Ishigaki *et al.* 2017[2]	72	R sides of vulva for 10 years	Invasive Ductal carcinoma	ER +, PR +, HER2 −	Lymph Nodes biopsy: SLN 0/1 Surgery: WLE Adjuvant therapy: Hormonal therapy, AI	Alive (6)
29	Sue Li *et al.* 2018[18]	69	4.6 cm mass on L vulva for 20 years, bleeding	Adenocarcinoma	ER +, PR +, HER2	Lymph Nodes biopsy: N/A Surgery: Radical vulvectomy, Adjuvant therapy: AI (Anastrazole 1 mg orally daily)	Alive (18)
30	Ananthula *et al.* 2020[1]	47	Painful 2.7 cm nodule for 1 year, Swelling, bleeding	Adenocarcinoma	GATA 3	Lymph Nodes biopsy: 4/17 Surgery: Radical vulvectomy, bilateral LND Adjuvant therapy: Tamoxifen, CDK4/6	DOD
31	Farrag *et al.* 2020[3]	53	(2 × 2) cm nodule on L labia for 2 years, discomfort	DCIS	ER +, PR −, HER2 −	Lymph Nodes biopsy: N/A Surgery: N/A Adjuvant therapy: Hormonal therapy (Tamoxifen)	Alive (36)

Abd, abdomen; AI, aromatase Inhibitors; DOD, dead of disease; ER, oestrogen; F-U, follow-up; L, left; LND, lymph node dissection; N/A, not applicable; PR, progestin; PY, publication year; R, right; RT, radiation therapy; WLE, wide local Excision.
[a]cases that had co-occurrence with breast cancer.

mammography, computed tomography of the chest, pelvis, and abdomen, magnetic resonance imaging of the upper abdomen, a colonoscopy, a bone scan, and tumour markers. If the in-situ component cannot be detected, and there is no concurrent breast cancer, the definitive diagnosis relies on identifying morphologic patterns consistent with breast carcinoma in addition to the presence of oestrogen receptors and/or progesterone receptors and/or the presence of common breast cancer markers such as epithelial membrane antigen, carcinoembryonic antigen, and glandular keratins[19]. Primary EBCs of the vulva are staged according to the tumour, node, and metastasis system that applies to breast cancer[1].

Differential diagnoses include extramammary Paget disease, adenocarcinomas arising from the Bartholin glands, sweat gland carcinomas, and metastatic cancer due to their similar histopathological patterns[20]. Due to the rarity of the disease, there are no current guidelines for the management of primary ductal adenocarcinoma of mammary-like glands of the vulva[18]. As a result, recommendations are to follow the principles used to treat orthotopic breast cancer of the same stage[2,17,20,21]; hence, the management is mostly surgical, followed by an individualized combination of radiotherapy, chemotherapy, and/or hormonal therapy[1,20]. The surgical management of the EBC of the vulva is variable and ranges from local excision to bilateral radical vulvectomy. Of the 31 cases of primary ductal adenocarcinoma of mammary-like glands of the vulva reported in the literature, 11 patients underwent bilateral radical vulvectomy[1,8,10-12,17,18,21-24]. In addition, lymphadenectomy was performed in cases where lymph nodes were affected[1,8,11,12,15,17,19,21–29]. Sentinel lymph node mapping in vulvar cancer is a contemporary topic in the medical literature that has been reported in only a few cases in the literature and is suggested to be a part of the standard treatment for selected patients in the early stages of vulvar cancer[2]. However, its role remains unclear[18].

Regarding postoperative treatment, 23 of all the reported cases of primary ductal adenocarcinoma of the vulva had positive oestrogen and/or progesterone receptors; of these, 12 were treated with Tamoxifen postoperatively, whereas the remaining cases were treated based on the individual status[1,3,8,11,17,19,22,25,26,29–31]. The EBC follow-up data are limited due to the rarity of the disease, and thus its prognosis remains unclear[2]. The survival rate could range between 4 months and 3 years. Nine of these cases were alive at around 2-year follow-ups[8,15,18,20,21,29–32] and two cases were alive at 3 years[3,19] (Table 1).

Conclusions

EBC of the vulva is rare, and only a few cases are reported in the literature. It is crucial to differentiate primary breast cancer of the vulva from metastatic cancer in this area. Therefore, a thorough history, physical examination, and diagnostic tests should be performed. Since there are no current guidelines on the treatment of such cases, the treatment follows the guidelines for treating orthotopic breast cancer. The report of similar cases in the literature would be helpful to establish a database and develop guidelines for the diagnosis and management of primary ductal adenocarcinoma of the mammary-like glands of the vulva and primary EBC in general.

Ethical approval

Ethics approval is not required for case reports at our institution. Institution name: Obstetrics University Hospital, Damascus, Syrian Arab Republic.

Consent

A consent was obtained from the patient for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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Author contribution

All authors read and approved the final manuscript. M.M.: design of the study, data collection, data interpretation and analysis, drafting, critical revision, approval of the final manuscript. O.Z.: data collection, data interpretation, and analysis, critical revision, drafting, approval of the final manuscript. D.N.: data interpretation and analysis, critical revision, drafting, approval of the final manuscript. A.A.: drafting, critical revision, approval of the final manuscript. M.C.: drafting, critical revision, approval of the final manuscript. W.RA.: drafting, critical revision, approval of the final manuscript. D.Y.: drafting, critical revision, approval of the final manuscript. I.K.: drafting, critical revision, approval of the final manuscript. A.M.D.: preparing correspondence files, preparing the final manuscript, approval of the final manuscript. B.K.: The Supervisor, patient care, drafting, critical revision, approval of the final manuscript.

Conflicts of interest disclosure

The authors declare that they have no conflicts of interest.

Research registration unique identifying number (UIN)

1. Name of the registry: NA.
2. Unique identifying number or registration ID: NA.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): NA.

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Dr. Bashar Kurdi.

Data availability statement

Not applicable. All patient data generated during this study are included in this published article and its supplementary information files.

Provenance and peer review

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