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**Case Report**

# A rare presentation of familial polymastia with associated polycystic ovarian morphology: a case report with a one-year follow-up

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## ABSTRACT

Polymastia is a congenital anomaly resulting from incomplete embryological regression of the milk line. While relatively common, its familial occurrence and association with other morphological conditions present unique clinical scenarios. This report details a rare familial presentation of polymastia with concurrent polycystic ovarian morphology to highlight the need for holistic diagnostic evaluation. An 18-year-old female with a positive family history of polythelia presented with a left anterior axillary mass. Breast ultrasonography confirmed an accessory nipple with a distinct ductal system suggestive of left-sided polymastia. Abdominopelvic ultrasonography revealed bulky ovaries with tiny cysts (right ovary volume 6.61 CC, left ovary 9.69 CC), indicating polycystic ovarian morphology. The patient underwent complete surgical excision without complications. Histopathology demonstrated benign squamous epithelium with slight hyperkeratosis, pilosebaceous structures, smooth muscle, and breast lobules with ducts in the deep dermis, confirming no evidence of malignancy. At the one-year follow-up, the patient remained entirely asymptomatic with no recurrence and a satisfactory cosmetic outcome. This case emphasizes the clinical necessity of comprehensive systemic evaluation in patients with congenital anomalies. Surgical excision remains the definitive management for polymastia, successfully addressing cosmetic concerns and ruling out malignant changes.

**Keywords:** Accessory breast, Case report, Polymastia, Polycystic ovary syndrome, Supernumerary nipple

## INTRODUCTION

Accessory breast tissue refers to residual breast tissue resulting from the failure of regression during embryogenesis.[1,2] Ectopic breast tissue acts as an umbrella term for both aberrant and supernumerary tissue, which are most frequently found along the "milk line" extending from the anterior axillary fold to the medial aspect of the thigh.[3,4] While these cases are most commonly sporadic, familial inheritance patterns with incomplete penetrance have been documented in clinical literature.[5,6]

This ectopic breast tissue is subject to physiological and pathological changes identical to normal breast tissue, including benign cystic changes, fibroadenomas, and, rarely, carcinomas.[7,8] Patients frequently present to clinicians due to cosmetic disfigurement, severe anxiety, or premenstrual pain caused by increased vascularity in the accessory tissue.[9]

Furthermore, supernumerary nipples and accessory breasts have been historically classified by Kajava into eight distinct anatomical classes, a system that remains widely utilized in modern clinical evaluation.[10] We present a rare case of an 18-year-old female presenting with left-sided polymastia, a documented positive family history of polythelia, and concurrent ultrasonographic findings of polycystic ovaries, managed successfully with targeted surgical excision.

## CASE REPORT

An 18-year-old female presented to outpatient department with a chief complaint of persistent swelling and an extra nipple located just above and slightly lateral to the orthotopic nipple. Physical examination confirmed the presence of left-sided polymastia (a complete accessory breast including glandular tissue, areola, and nipple). The overlying skin was grossly unremarkable (Figure 1).

Of significant clinical note, the patient possessed a positive family history of polythelia, which was specifically documented in her maternal cousin's daughter. Targeted breast USG of the left breast demonstrated accessory glandular tissue with a nipple connected to its own distinct ductal system. No solid or cystic mass lesions, calcifications, or dilatation were noted, thereby confirming the clinical diagnosis of polymastia.[11] To evaluate the extent of the anomaly and screen for any associated pelvic pathologies, ultrasonography (USG) of the abdomen and pelvis was performed. The abdominopelvic ultrasonography (USG) revealed bulky ovaries showing multiple tiny cysts, suggestive of underlying polycystic ovarian disease (PCOD).

Following standard preoperative evaluation and after obtaining informed written consent, the patient underwent surgical management. Complete surgical excision of the polymastic tissue was performed uneventfully (Figure 2 and 3).

The completely excised specimen was sent for formal histopathological examination, which revealed tissue lined by benign squamous epithelium exhibiting slight hyperkeratosis. Pilosebaceous structures, smooth muscle, and breast lobules with distinct ducts were clearly identified in the deeper dermis, consistent with accessory breast tissue, and absolutely no evidence of malignancy (Figure 4 A and B, Figure 5 A and B).

The postoperative recovery period was entirely uneventful. At the formal one-year follow-up examination, the patient remained completely asymptomatic. There was no clinical or radiological evidence of local recurrence, and the patient reported a highly satisfactory cosmetic outcome (Figure 6).

**Table 1: Kajava classification of supernumerary breast tissue.**

| Class[a] | Terminology | Tissue components present |
|---|---|---|
| **Class I** | Polymastia | Complete breast(s) with nipple, areola, and glandular tissue |
| **Class II** | Supernumerary breast without areola | Nipple and glandular tissue, no areola |
| **Class III** | Supernumerary breast without nipple | Areola and glandular tissue, no nipple |
| **Class IV** | Mamma aberrata | Only glandular tissue |
| **Class V** | Pseudomamma | Nipple and areola, no glandular tissue, replaced by fat |
| **Class VI** | Polythelia | Nipple only |
| **Class VII** | Polythelia areolaris | Areola only |
| **Class VIII** | Polythelia pilosa | Patch of hair only |

[a]Classification system established by Yrjö Kajava in 1915.

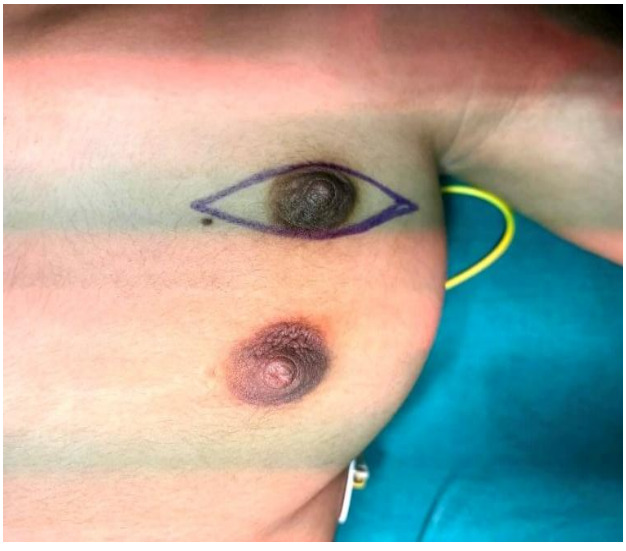

**Figure 1: Gross clinical presentation of left breast showing polymastic tissue along the anterior axillary line.**

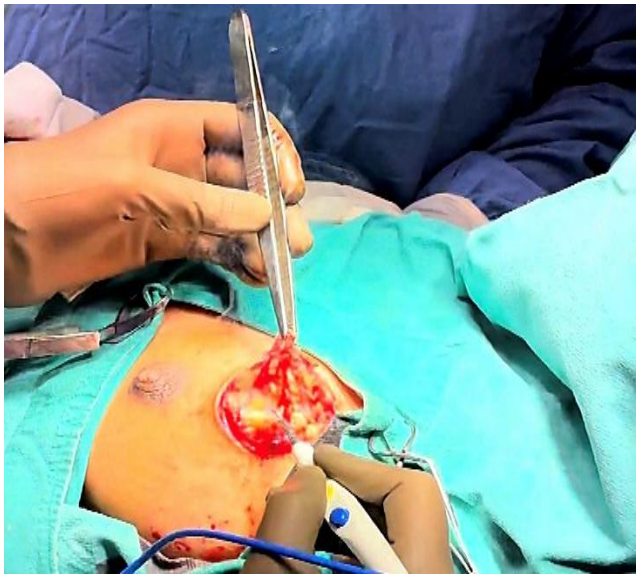

**Figure 2: Intra-operative image showing surgical excision of accessory breast tissue.**

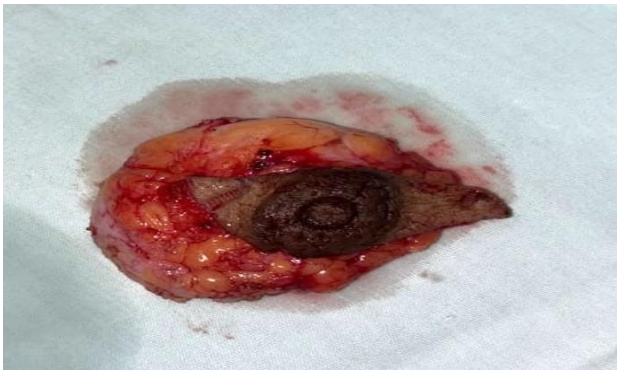

**Figure 3: The surgically excised tissue specimen.**

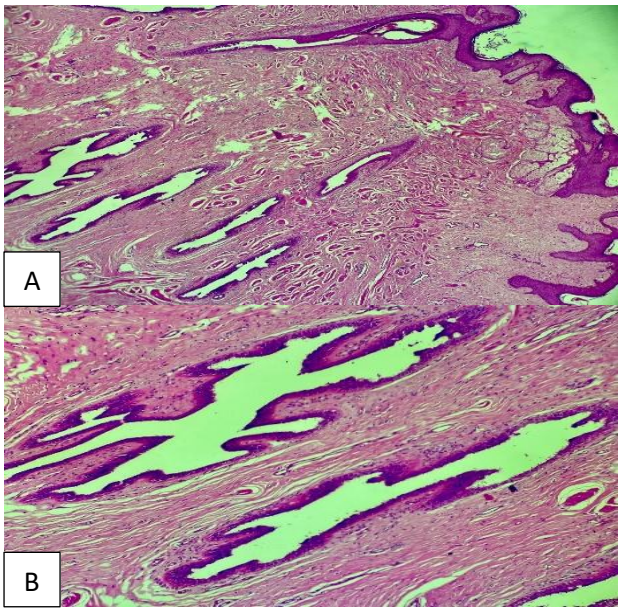


**Figure 4 (A and B): Histopathological examination. (A) Benign squamous epithelium with slight hyperkeratosis and underlying dermis. (B) Pilosebaceous structures and smooth muscle.**

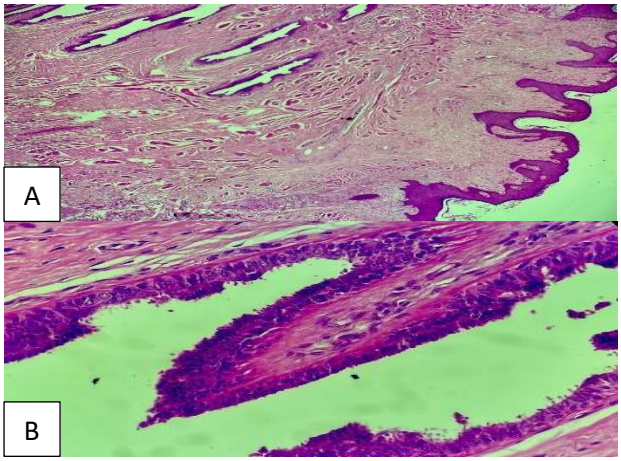


**Figure 5 (A and B): Histopathological examination. (A) Breast lobules with distinct ducts in the deep dermis. (B) High-power view confirming the absence of cellular atypia or malignancy.**

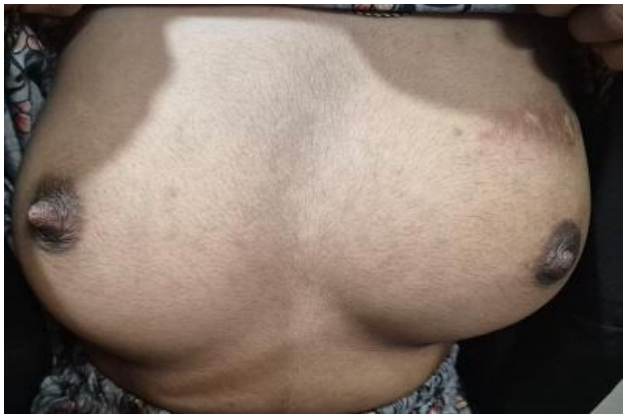

**Figure 6: Clinical appearance at the one-year postoperative follow-up, demonstrating a satisfactory cosmetic outcome with no recurrence.**

## DISCUSSION

Accessory breast tissue fundamentally results from the failure of the embryological mammary ridges (milk lines) to fully regress during normal fetal development.[2,12,13] While it can occur anywhere along this anatomical line, the axillary region remains the most common clinical presentation.[3,4] In 1915, Kajava established a comprehensive classification system for supernumerary breast tissue based heavily on the presence of the nipple, areola, and functional glandular tissue (Table 1), which remains the standard clinical taxonomy utilized globally today.[10]

According to this rigorous classification, our patient's specific presentation strictly aligns with class I (Polymastia) as complete glandular tissue, an areola, and a nipple were clinically and sonographically evident, though the ductal system was isolated.[10,14]

A highly notable aspect of this case is the documented positive family history of polythelia in the patient's cousin's daughter. While the vast majority of cases are entirely sporadic, familial inheritance exhibiting an autosomal dominant pattern with incomplete penetrance has been previously described in the literature, emphasizing the genetic underpinnings of mammary ridge regression failures.[5,6]

Furthermore, the incidental finding of polycystic ovarian morphology (bulky ovaries with tiny cysts and a high AF score) highlights the absolute clinical necessity for comprehensive systemic evaluation in patients presenting with congenital anomalies. While the direct genetic or endocrinological link between PCOD and polymastia requires further large-scale epidemiological investigation, the concurrent presentation in this young female underscores the vital importance of a holistic diagnostic approach. Surgical excision remains the definitive gold standard of treatment, effectively addressing patient cosmetic concerns, alleviating symptoms like premenstrual pain, and definitively ruling out

malignancies such as carcinomas or fibroadenomas that can, albeit rarely, arise in ectopic breast tissue.[9,14,15]

## CONCLUSION

Familial polymastia, particularly when presenting as a complete accessory breast (Kajava Class I), is a rare clinical entity that necessitates a comprehensive diagnostic approach, especially when concurrent with morphological anomalies such as polycystic ovarian syndrome. As evidenced by our gross clinical imaging targeted surgical management remains the optimal strategy. Complete surgical excision not only provides significant symptomatic relief and a satisfactory cosmetic outcome but also definitively eliminates the inherent risk of future malignant transformation within the ectopic breast tissue.

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