PJSS Vol. 74, No. 2, July-December, 2019

PJSS PHILIPPINE JOURNAL OF SURGICAL SPECIALTIES

Adult-Onset Cystic Hygroma in the Axilla in a 44-year old Female: A Case Report

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This is a case of a 44- year old female presenting with an 18cm x 17cm soft, movable, non-tender mass at the right axilla extending to the lateral aspect of the right breast. Computerized tomographic scan of the chest revealed a lobulated, multi-septated hypodense mass. The patient underwent excision of the right axillary mass and final histopathology revealed cystic lymphangioma. Adult-onset cystic hygroma of the axilla is a rare case, with less than 10 studies documented in PubMed. Total surgical excision remains to be its primary treatment.

Keywords: Adult-onset cystic hygroma, cystic hygroma, lymphangioma, axilla

Cystic Hygroma, also known as lymphangioma, is a benign congenital malformation of the lymphatic system that is caused by an obstruction of communication between the lymphatic and venous systems.[1,2] The lack of communication results to lymphatic accumulation in the areas of major lymphatic channels, especially at areas with less resistance, including the cervical and the axillary regions.[3,4] Eighty percent of cystic hygromas occur in the neck, usually at the posterior cervical triangle. The axilla, superior mediastinum, mesentery, retroperitoneal region, pelvis, and lower limbs are also areas of occurrence.[1]

Cystic hygroma is typically found in children and usually occurring in the cervico-facial region.[5,6,7] Its diagnosis is uncommon in adults and literature review has shown rare data on adult- onset cystic hygroma in the axilla.[5,6,8]

The main objective of this paper was to present a rare case of adult-onset cystic hygroma of the axilla, and to report known data of the case based on published medical literature, reviewing its clinical presentation, diagnosis, pathologic findings and management.

The Case

The patient is a 44-year-old, single, unemployed, Filipino female with a five-year history of progressively enlarging, soft, movable, non-tender right axillary mass. There was no history of trauma, infection, or surgery at the affected site. No previous consults were done and no medications were taken for the mass.

Patient is a known hypertensive for 17 years and has been taking oral Amlodipine 5 mg daily, taken with good compliance. No herbal medications, no contraceptive pills and no hormonal replacements were taken. She has no other co-morbidities. Patient has no known food and drug allergies. She is a non-smoker, a non-alcoholic beverage-drinker, and has no history of illicit drug use. Patient's only previous surgery was repair of fourth degree perineal laceration due to childbirth. Patient's only known heredo-familial disease is hypertension on the maternal side and has no family history of any congenital abnormalities.

On physical examination, the patient had a sthenic habitus, with a height of 145 cm and weight of 50 kg. The patient's BMI is 23.7 kg/m^2. The patient had normal vital signs and cardiopulmonary exam findings. An 18cm x 17cm soft, movable, non-tender mass was noted at the right axilla extending to the lateral aspect of the right breast (Figures 1 & 2). There was no skin dimpling, no nipple discharge, nor palpable lymph nodes at the right axilla, supraclavicular

and infra-clavicular areas. The right breast also had a 2cm x 3cm firm movable, non-tender mass at the 12 o'clock position and a 2cm x 2cm firm, movable, non-tender mass at the 7 o'clock position. The left breast was noted to have a 2.4cm x 1.4cm firm, movable, non-tender mass at the 6 o'clock position.

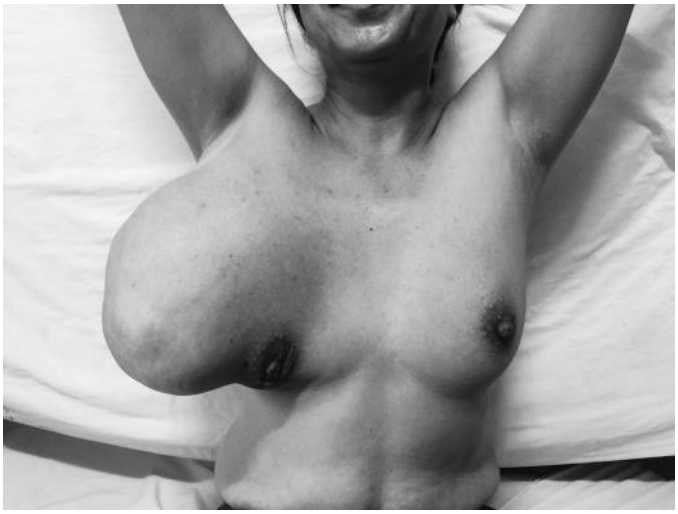

Figure 1. Patient's axillary mass (frontal view)

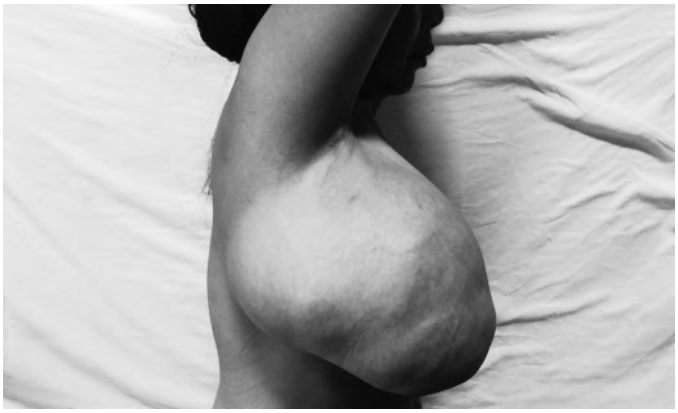

Figure 2. Patient's axillary mass (lateral view)

Ultrasonographic examination of the right axilla showed a huge multiseptated cystic mass at the right axilla. CT scan of the chest was done which revealed a lobulated multi-septated hypodense mass measuring 16.5cm x 16.4cm x 17cm at the right axillary region extending to the right subpectoralis and right anterolateral chest wall/breast (Figure 3). There were well-defined soft tissue nodules at the upper inner quadrant measuring 2.0cm x 1.4cm and at the outer mid half of the right breast measuring 2.1cm x 1.3cm. A soft tissue nodule at the retroglandular fat of the lower outer quadrant of the left breast was also noted. No pre-operative biopsy was taken. Our pre-operative impression was 1) lymphangioma of the right axilla, 2) fibroadenomata of both breasts.

The patient underwent excision of adult axillary cystic hygroma of the right axilla and excision of fibroadenomata of the right breast under general anesthesia. Pre-operative medications included: oral celecoxib 400 mg as pre-emptive analgesia, parenteral ranitidine 50 mg, and parenteral cefoxitin 2 grams as prophylaxis.

The patient was positioned supine. The right arm was placed over the head. The operative site was aseptically prepared including the right arm and forearm. Sterile drapes were placed accordingly to expose the right axilla and lateral side of the right breast (Figure 4). The right arm was held in place throughout the procedure. The patient was put under general anesthesia. A curvilinear incision was made around the mass and the incision was carried down to the subcutaneous tissue and up to the capsule of the mass (Figure 5). The mass was carefully dissected from the pectoralis major muscle posteriorly and from the anterior border of latissimus dorsi laterally using electrocautery and sharp and blunt dissection. The cystic hygroma was removed en bloc (Figures 6 & 7). Hemostasis was done. Excision of the fibroadenomata located at the 12 o'clock position and 7 o'clock positions of the right breast was done using a posterior approach through the same incision. Two Jackson-Pratt drains were inserted, one placed at the axillary area, and the other above the pectoralis major muscle. The subcutaneous tissue was closed using Polyglactin 910 4-0 sutures in an inverted simple interrupted manner and the skin was apposed with Polypropelene 3-0 sutures in a continuous subcuticular manner (Figure 8). The wound was dressed and the operation was concluded.

The patient had a follow-up visit 1 month post-operatively and had no complaints of pain, discomfort or numbness. The incision site had healed well and there was no limitation of movement on the right upper extremity (Figure 9).

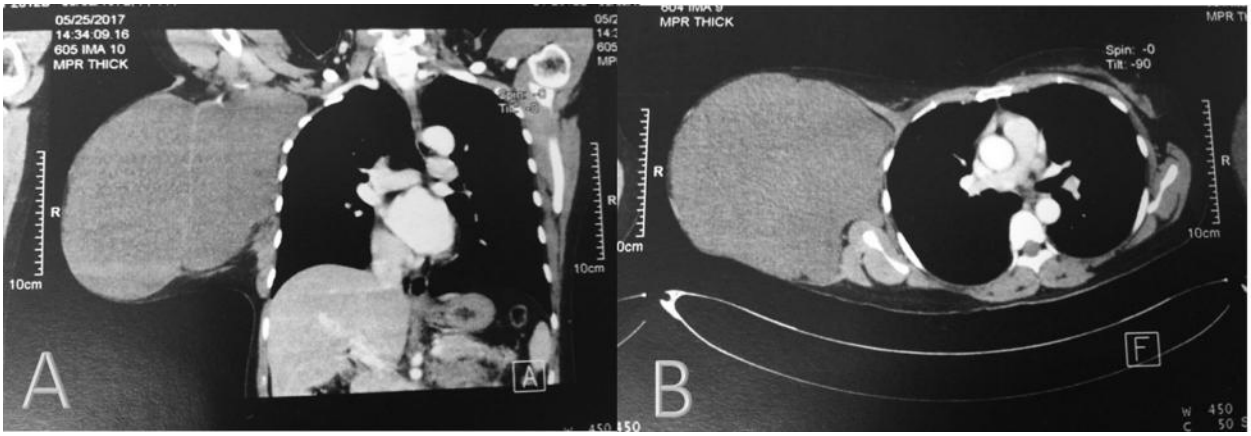

Figure 3. CT scan of the chest. A, Coronal view. B, Axial view.

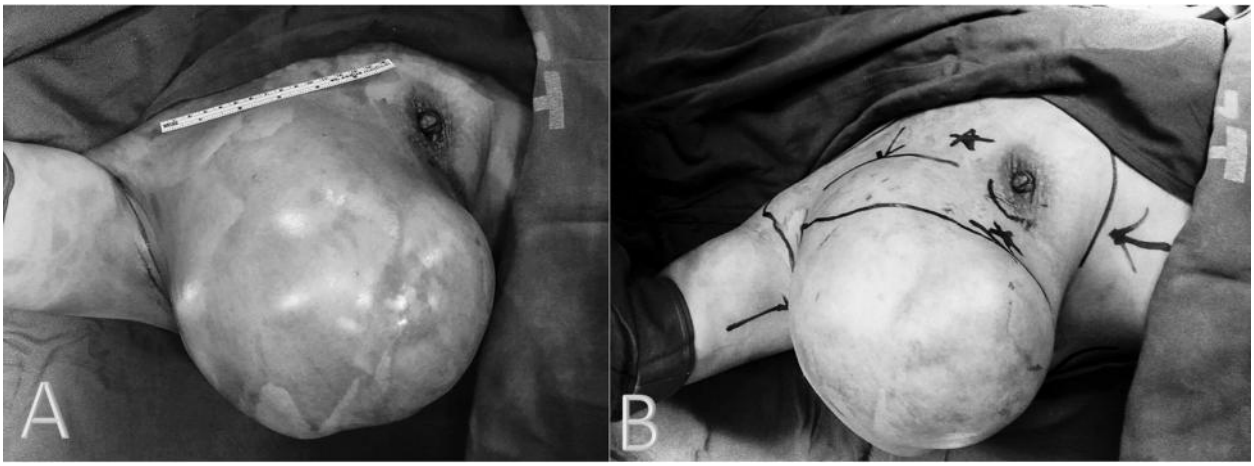

Figure 4. Pre-operative photograph of the mass. A, unmarked. B, with marks for surgical planning.

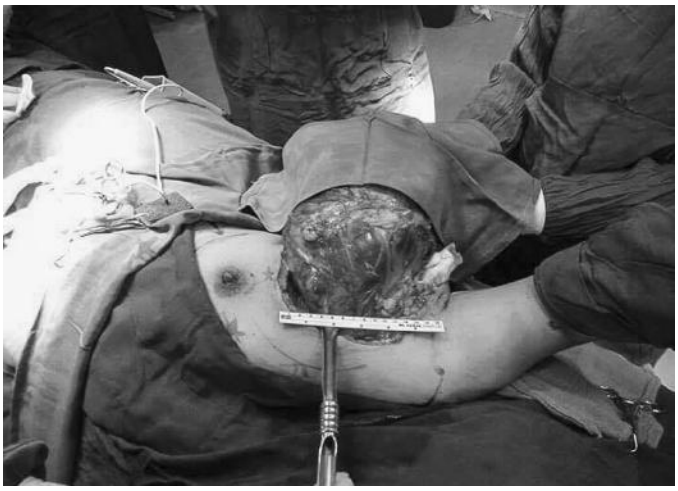

Figure 5. Patient's mass intraoperatively.

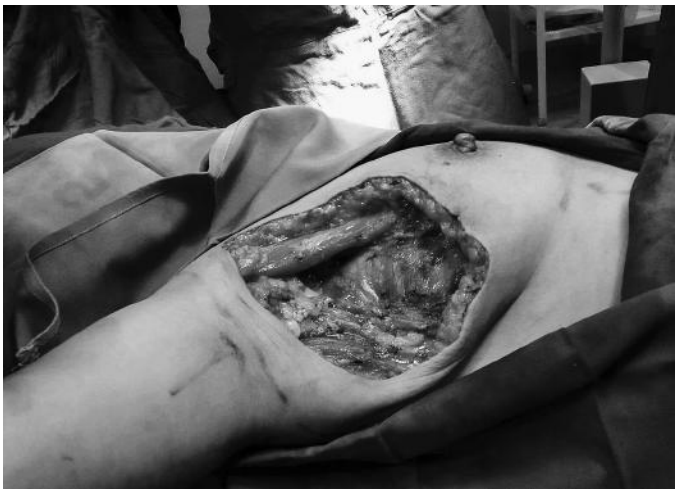

Figure 6. Defect of the operative site after removal of the mass.

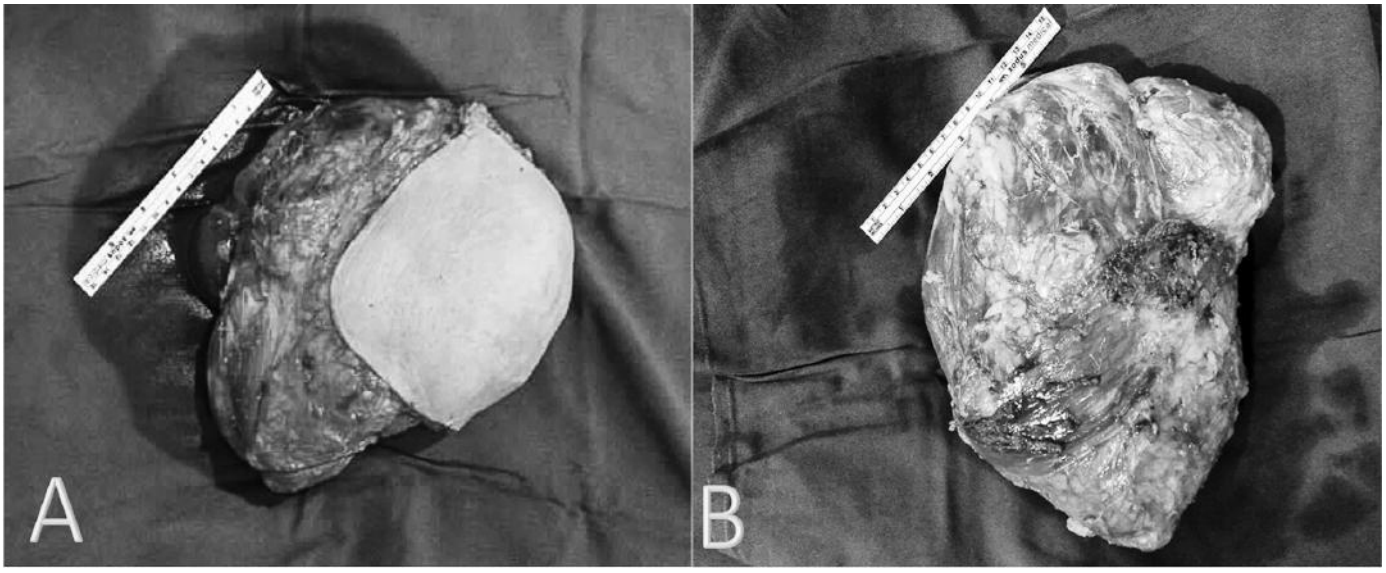

Figure 7. Axillary mass specimen consisting of a large intact fluctuant cyst measuring 18 cm at its widest dimension. A, anterior. B. posterior.

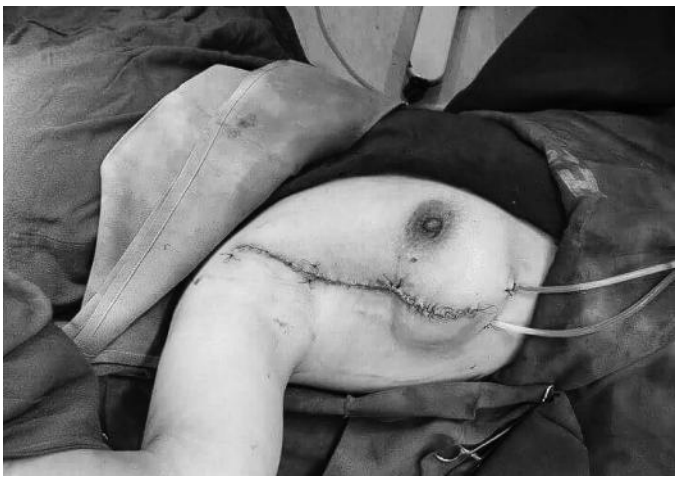

Figure 8. Post-operative site of the patient.

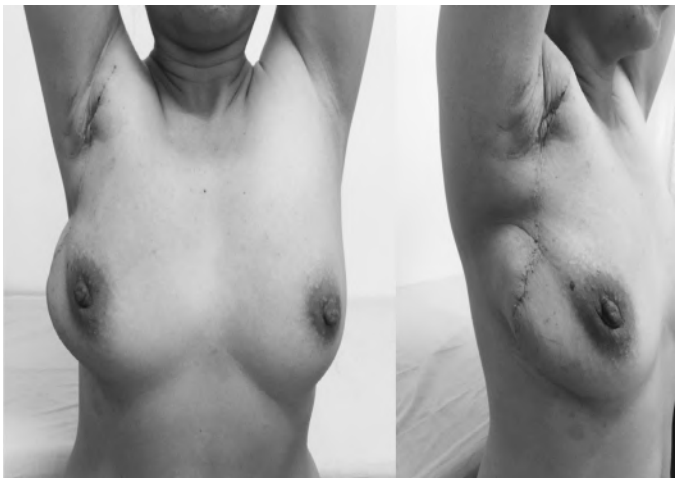

Figure 9. Photographs of patient one-month post-operation.

The final histopathology report of the right axillary mass revealed cystic lymphangioma (hygroma). The mass consists of a large intact fluctuant cyst, measuring 180mm at its widest dimension. This is covered on one side by an irregularly ellipsoid intact skin and fibrofatty tissue in the cyst external surface. Section shows a large locule containing abundant pale yellow clear fluid. The cyst internal surface has abundant trabeculations. A few smaller locules are also seen in some parts of the cyst wall (Figure 10). The right breast mass at the 12 o'clock position was read as Fibroadenoma measuring 20mm x 15mm x 11mm. The right breast mass at the 7 o'clock position measured 16mm x 14mm x 10mm and was read as Fibrocystic disease with fibrosis, microcysts, macrocysts and focal apocrine metaplasia. The final diagnosis for the axillary mass was adult cystic hygroma of the right axilla.

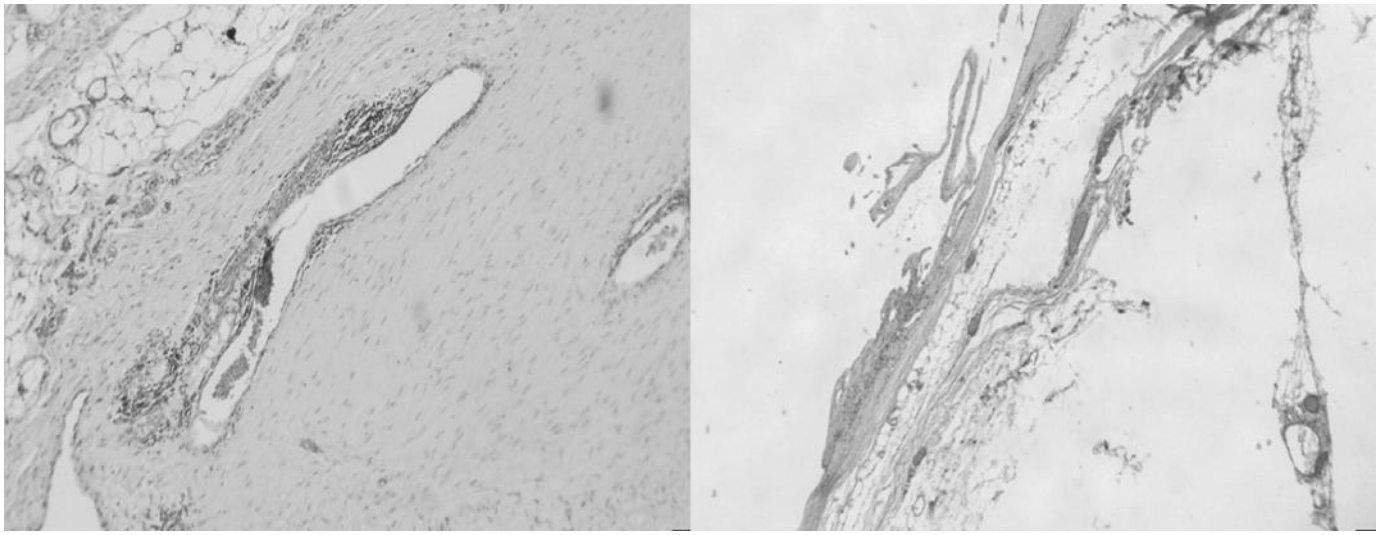

Figure 10. Histologic slides of the patient's mass on H & E stain shows sections from a cyst wall formed by fibromuscular tissue and internally lined by a layer of markedly attenuated flattened epithelium, consistent with cystic hygroma.

Discussion

Cystic Hygroma is a benign congenital anomaly of lymphatic origin, most often observed in infants and children.[5,6,10] Axillary cystic hygroma is rare in adults.[3,4,9,10,11] Its development in adulthood has been proposed to be as a result of late proliferation of cellular nests of the lymphatic system and to be related to predisposing factors such as trauma, infection, tumor growth or iatrogenic stimuli.[9] In the case presented, the patient had no documented predisposing factors such as history of trauma or infection at the affected site.

Cystic hygromas are mostly reported as progressive tumors.[3,9,10,11] On physical examination, cystic hygromas present as soft, painless masses. When located superficially, these masses can transilluminate.[1] Infection and hemorrhage can cause rapid expansion of the mass which may cause pain and discomfort.[1, 5] Possible complications of cystic hygromas include bleeding, infection, and fistula formation.[5] The patient in this study had a progressive presentation of axillary mass, which was soft and painless.

The evaluation of soft tissue tumors, including cystic hygromas is better done through Magnetic Resonance Imaging (MRI) as compared to Computerized Tomography (CT), since contrast between tumor and muscle and between tumor and vessels are better seen in MRI.[12] Vital neurovascular structures adjacent to the tumor are crucial in planning for surgical treatment, hence MRI is recommended for all patients.[13] Fine needle aspiration biopsies are often non-diagnostic since the architecture of the lesion cannot be evaluated.[6]

Recurrence is common with incompletely resected cystic hygromas.[1] In an emergency, aspiration may be done to provide temporary relief.[1] Interferon alpha, laser therapy and intralesional sclerosing agents are nonsurgical treatment options for cystic hygromas.[1,14] OK-432 (Picibanil), a streptococcal immunotherapeutic agent is the sclerosing agent of choice.[1] Bleomycin is a sclerosing agent that harbors the risk of pulmonary toxicity.[1]

Complete surgical excision is the treatment of choice for cystic hygromas, however its location may make an en bloc resection challenging.[5,14] Since cyst remnants cause tumor recurrence, total excision of the cyst affects prognosis.[14]

According to Riechelmann, et al. complete disease control for cystic hygroma can be achieved after total removal of the cystic structures; 89% disease control after subtotal removal; 14% disease control after partial removal; and 0% disease control after incision and aspiration of cystic hygroma.[15]

Acknowledgements

The authors would like to acknowledge the invaluable support of Dr. Saleshe Tracy Anne Baking-Fernandez for being instrumental in the surgical management of this case and Dr. Cherry Lyn Montealto for her assistance in making this report.

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