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Systematic Review: Littoral Cell Angioma of the Spleen

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Abstract: ***Background: Littoral Cell Angioma is a rare vascular tumor of spleen mostly diagnosed incidentally and though have malignant potential; may often run a benign course. Given its rarity, it is of utmost significance to throw more light onto the clinical features, diagnostic approaches, therapeutic interventions and outcomes associated with LCA which has been served through this systematic review of existing literatures. Method: A cumulative total of 215 records was identified via database searches. Data extraction was carried out and the quality of case reports and case series was evaluated using the Joanna Briggs Institute (JBI) critical appraisal tools. Following the elimination of duplicates and the screening of titles and abstracts, 34 studies qualified for comprehensive full-text evaluation. Finally, 21 studies, encompassing a total of 85 cases of Littoral Cell Angioma, satisfied the inclusion criteria and were incorporated into the systematic review and reported in accordance with the PRISMA guidelines. Results: Out of the 85 cases of LCA, it was found that there was a slight female preponderance with incidental detection predominating in majority of the cases. Total splenectomy was preferred against other surgical options. HPE and IHC forms the mainstay of the diagnosis. Overall prognosis is good except for rare malignant transformation. Discussion: This review highlights the need for heightened awareness of LCA among clinicians, especially when evaluating splenic masses. Further research is necessary to identify the molecular pathways underlying malignant transformation and to develop potential biomarkers for early detection.***

Keywords: Littoral Cell Angioma, Littoral cell tumors, Spleen neoplasm and vascular splenic tumor

1.Introduction

Littoral cell Angioma [LCA], is a rare splenic vascular neoplasm that originates from littoral cells lining the sinusoids of the red pulp. These tumors have both benign and malignant potential. Thus, it is intricate to assess their clinical importance. The literature available on LCA has mainly been case reports and sparse case series due to its rarity. This systematic review aims to summarize the clinical features, diagnostic approaches, therapeutic interventions and outcomes associated with LCA.

Objective

On the basis of literature obtained thus far, we aim to critically and comprehensively evaluate the clinical presentation, methods of diagnosis, treatment strategies and outcomes of patients diagnosed with Littoral cell angioma of the spleen.

Methodology

Search Strategy

Searches were conducted in major medical databases, including PubMed, Scopus, and Web of Science, from their inception until August 2024. "Littoral Cell Angioma", “Littoral cell tumors”, “Spleen neoplasm" and "Vascular splenic tumor" were used as keywords.

Eligibility Criteria

Inclusion Criteria:

- Studies reporting confirmed cases of Littoral Cell Angioma of the spleen.
- Case reports, case series, and retrospective studies.
- Articles on the diagnostic methods, clinical features, therapeutic interventions, and outcomes.

Exclusion criteria:

- Studies focusing on other types of splenic vascular tumors.
- Studies not reported in English language
- Studies with no confirmed pathological diagnosis of LCA.

Study Selection Process

Two independent evaluators conducted a screening of all titles and abstracts to determine potential relevance. The complete texts of studies deemed eligible were evaluated by the same reviewers, and any inconsistencies were reconciled through consensus.

Data Extraction

Data extraction was carried out using a pre-defined template. Extracted information included:

- Demographic details (age, gender)
- Clinical presentation
- Diagnostic modalities (e.g., imaging, histopathology)
- Treatment approaches (e.g., splenectomy, chemotherapy)
- Outcomes (recurrence, metastasis, survival)

Quality Assessment

The quality of case reports and case series was evaluated using the Joanna Briggs Institute (JBI) critical appraisal tools. Each study was scored and only studies with a

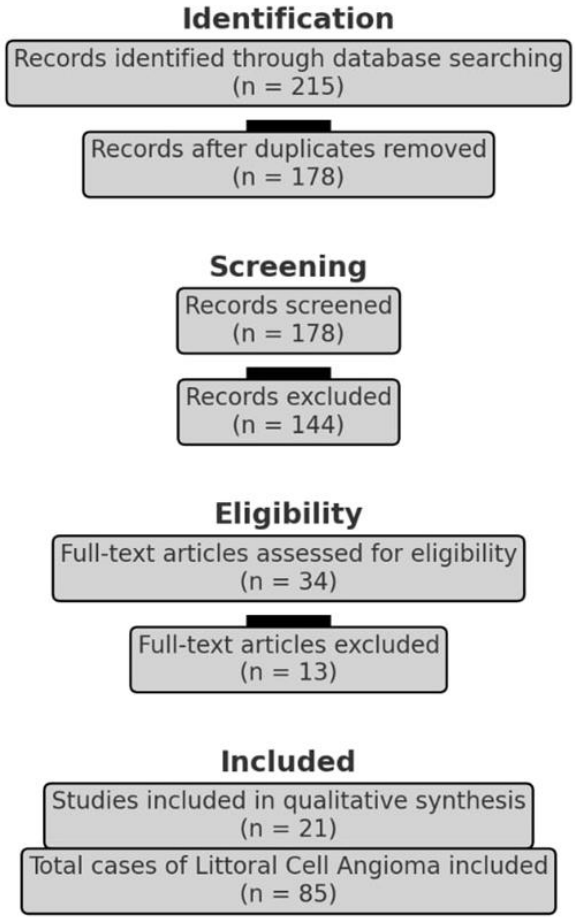
moderate to high quality were considered for the final analysis.

2.Results

Study Selection

A cumulative total of 215 records was identified via database searches. Following the elimination of duplicates and the screening of titles and abstracts, 34 studies qualified for comprehensive full-text evaluation. Finally, 21 studies, encompassing a total of 85 cases of Littoral Cell Angioma, satisfied the inclusion criteria and were incorporated into the systematic review.

PRISMA Flow Diagram:



Demographic Data

Age Range: 15 to 72 years

Gender Distribution: Slight female predominance (56%)

Clinical Presentation

Asymptomatic: 55% of patients were incidentally diagnosed during imaging studies for unrelated conditions.

Symptomatic: Abdominal pain was the most common symptom encountered, accounting for 32%; splenomegaly, for 20%; and weight loss, for 15%. Less frequently reported was hypersplenism or hemolytic anemia.

Diagnostic Modalities

Imaging:

Ultrasound and computed tomography (CT) were the most common initial diagnostic tools. LCA often presented as hypodense or isodense splenic masses on CT.

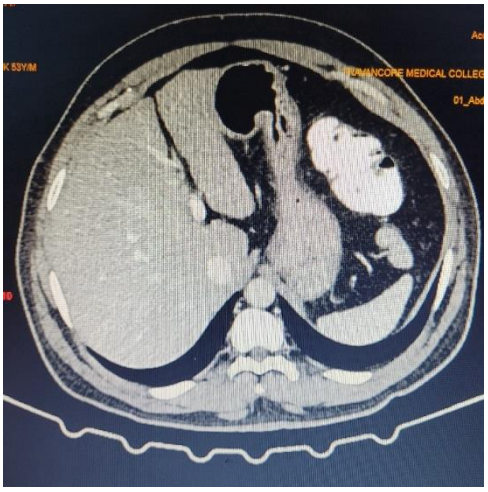


Figure 1: CT image of patient of the author showing tumor arising from the lower pole of spleen

Magnetic resonance imaging (MRI) was performed in some cases, showing varied signal intensities due to the hemorrhagic and cystic components of the tumor.

Histopathology:

Diagnosis was confirmed post-splenectomy via histological examination. Littoral cell Angiomas demonstrated positive immunohistochemical staining for markers such as CD31, CD68, and FVIII-related antigen. CD8 staining was consistently negative.

Treatment Approaches

Splenectomy: Total splenectomy was the definitive treatment in all cases where LCA was identified. No cases of partial splenectomy or non-surgical management were reported.

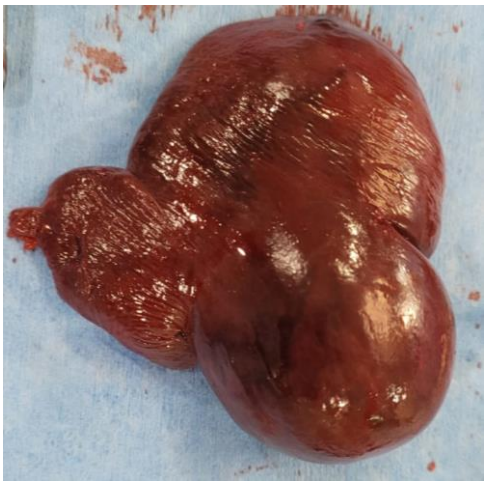


Figure 2: Gross specimen after total splenectomy in one of the author's patients

Indications: In symptomatic patients, splenectomy was indicated due to mass effect or suspicion of malignancy. For asymptomatic patients, splenectomy was often performed due to diagnostic uncertainty or concerns of progression to malignancy.

Follow-up:

In the majority of cases (92%), splenectomy led to complete resolution of symptoms with no recurrence reported on follow-up (mean follow-up duration: 24 months).

Malignant Transformation: Two cases of malignant LCA were reported. These were characterized by local recurrence post-splenectomy and required adjuvant chemotherapy.

Outcomes

Benign LCA: The prognosis for patients with benign littoral cell angioma was excellent, with no recurrence or metastasis in patients who underwent complete splenectomy.

Malignant LCA: The two patients with malignant transformation had poor outcomes, with one patient succumbing to metastatic disease within two years of diagnosis.

3.Discussion

This systematic review points out that LCA, despite its rarity, is mostly a benign condition and is often found incidentally. Histopathology is indispensable for establishing the diagnosis post-splenectomy since imaging characteristics are non-specific. Most of the patients have a good prognosis after splenectomy, while clinicians must be cognizant of the rare subset of cases with malignant potential.

The review underscores the need for heightened awareness of LCA among clinicians, particularly when evaluating splenic masses. Further research is necessary to delineate the molecular pathways underlying malignant transformation and to develop potential biomarkers for early detection.

4.Conclusion

Littoral cell Angioma of the spleen is a rare but largely benign vascular tumor. Splenectomy remains the gold standard treatment, with excellent outcomes in most patients. However, vigilant follow-up is essential to identify the rare instances of malignant transformation.

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