soho2023
ELEVENTH ANNUAL MEETING

UNIVERSITATEA DE STAT DE MEDICINĂ ȘI FARMACIE „NICOLAE TESTEMIȚANU” DIN REPUBLICA MOLDOVA

UNUSUAL PRESENTATION OF CASTLEMAN'S DISEASE IN PEDIATRIC PATIENT

Golban Rodica[2], Tomacinschii Cristina[1,3]

1. Nicolae Testemitanu State University of Medicine and Pharmacy, Chisinau, Republic of Moldova
2. Department of Hematology, Institute of Oncology of Republic of Moldova, Chisinau, Republic of Moldova
3. Institute of Mother and Child, Chisinau, Republic of Moldova

INTRODUCTION

Castleman's disease (CD) is a lymph node hyperplasia that can be unicentric or multicentric. In childhood, most patients have unicentric CD, which is typically curative with resection. Pediatric multicentric CD is rarely observed and requires systemic therapy.

Paraneoplastic pemphigus (PNP) is a mucocutaneous disorder characterized by mucocutaneous manifestations associated with an underlying tumor that is usually suspected because of resistance to treatment.

The clinical picture of PNP is characterized by polymorphic mucocutaneous manifestations with intractable stomatitis as the most characteristic clinical sign but may also be present conjunctivitis, genital ulcers, skin eruption, or it may mimic other pathologies such as herpes simplex virus infection, or other pemphigus.

No formal diagnostic or treatment protocols exist for PNP.

AIM

Evaluation of a case report of pediatric CD and its diagnostic and therapeutic management.

RESULTS

A 13-year-old Caucasian girl, with no family history of inflammatory diseases or malignancies, was completely healthy until the installation of a dental appliance, which was followed by the appearance of painful lingual, oral, and lip erosions.

Despite various therapies, including antiseptics, antibiotics, and oral corticosteroids, the oral symptoms recurred.

Due to concerns about paraneoplastic manifestations, she was investigated for underlying malignancy at another medical center using PET-CT, which showed an avid lesion in the left iliac fossa (45 mm). The patient was referred to another medical center where a needle biopsy was performed, revealing non-specific lymphoid tissue with fibro-sclerotic changes.

The next step was an exploratory laparotomy, and histology revealed the hyaline vascular variant of Castleman's disease. A PET scan showed retroperitoneal lymph nodes (4 × 3 × 6 cm, SUV 4.5), confirming the diagnosis of multicentric appearance of CD. Rituximab was initiated at a dose of 375 mg/m2/week for 4 consecutive weeks, but unfortunately, the response was unsatisfactory, with only a mild reduction in lymph nodes and oral erosions.

Off-label therapy with Siltuximab was recommended.





Figure 1. Lingual and lip eosions (1,2), erosive mucositis (1)

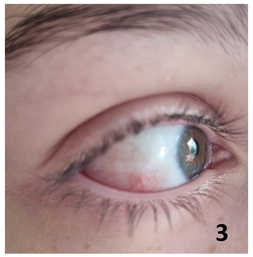






Figure 2. Conjunctival hyperemia (3) Skin vesicle on arm (4) Crusted lesions (5)

CONCLUSIONS

CD rarely occurs in childhood and remains a diagnostic challenge. Due to the paucity of cases there are no current guidelines for the selection of optimal therapeutic options and outcomes in pediatric cases, highlighting the need for further studies.

REFERENCES

- Talat N, Belgaumkar AP, Schulte KM: Surgery in Castleman's disease: a systematic review of 404 published cases. Ann Surg. 2012, 255:677-84.
- Kershenovich R, Hodak E, Mimouni D: Diagnosis and classification of pemphigus and bullous pemphigoid . Autoimmun Rev. 2014, 13:477-81.

CONTACT INFORMATION

Cristina Tomacinschii
Email:
cristinatomacinschii@gmail.com.