International Research Journal of Pharmacy and Medical Sciences
ISSN (Online): 2581-3277

Malignant Varicella in an Immunocompetent Patient: A New Case Report

R. Bouagaga[1], H. Bennasser Alaoui[2], H. Bachir[2], S. Hamaz[2], K. Serraj[2]
[1]Department of internal medicine, CHR Moulay Ali Chérif, Errachidia – Morocco
[2]Department of Internal Medicine, CHU Mohammed VI, Laboratory of Immuno-Hematology and Cellular Therapy, Faculty of Medicine and Pharmacy, Mohamed Premier University, Oujda. Morocco
E-mail: rachi90da@gmail.com

***Abstract*—** *Varicella is a highly contagious viral infection caused by the varicella zoster virus (VZV) and characterized by fever and a pruritic papulovesicular rash. In adults, its evolution can be marked by complications. We report a case of malignant varicella in an immunocompetent patient complicated by hypoxemic pneumonitis and thrombocytopenia. Under antiviral treatment; the evolution was favorable*.

***Keywords*—** *Varicella; immunocompetent; varicella pneumonitis; aciclovir*.

I. INTRODUCTION

Varicella is classically described as a highly contagious and almost "obligatory" viral disease of childhood, caused by the varicella zoster virus (VZV) and characterized by fever and an itchy papulovesicular rash [1]. It is considered to be a usually mild infection in children, whereas it can lead to fatal complications in adults [2]. Severe forms are often associated with immunodepression [3]. We report a case of malignant varicella in an immunocompetent adult; complicated by pneumopathy and thrombocytopenia.

II. OBSERVATION

The patient was 19 years old, the second of four siblings, with a sister who died of acute leukemia two years ago. The patient came to the emergency department with febrile dyspnea and a productive cough associated with a generalized rash that had been evolving for three days.

A careful clinical examination found a conscious, polypneic patient with a saturation of 78% on room air and 92% on 3L of O_2, tachycardia with a heart rate of 109 beats per minute, blood pressure of 120/60mmHg; with the presence of a pruritic, vesicular and crusted rash in places, with elements of different ages, on the face, trunk and limbs (figure 1). In addition, other purpuric necrotic lesions were found on the face and lower limbs (Figure 2). Crackling rales were auscultated on pleuropulmonary examination.

The initial laboratory work-up showed an inflammatory syndrome with a hyperleukocytosis of 16700/mm³ (predominantly lymphocytic); with a CRP of 176mg/l and thrombocytopenia of 18000/mm³. The rest of the work-up was unremarkable.

The thoracic angioscan showed bilateral micronodular lesions and condensations predominantly at the bases.

The diagnosis of malignant varicella was suspected in view of the typical appearance of the lesions and the evocation of a febrile pruritic vesicular rash in the two younger brothers a few days before the onset of the symptomatology.

HIV serology and QuantiFERON were negative; so were AAN and ANCA. A sternal puncture was normal.

The patient was put on oxygen therapy (3L of O_2 to the glasses), Aciclovir (10mg/Kg/8hours) and an antibiotic therapy (Amoxicillin-Ac clavulanic). The evolution was favorable; clinically, marked by apyrexia, regression of skin lesions, Oxygen withdrawal from the 5th day; and biologically (negative CRP and increased platelet count to 96,000/mm³). The duration of hospitalization was 12 days. Varicella serology, recovered one week after discharge, was positive.

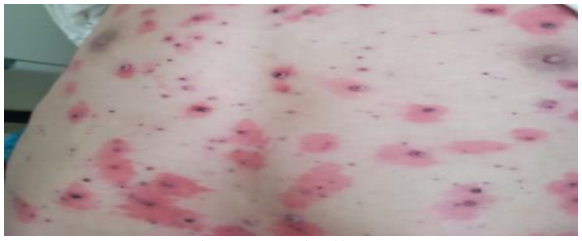

Figure 1: Lesions made of elements of different ages including umbilical vesicles.

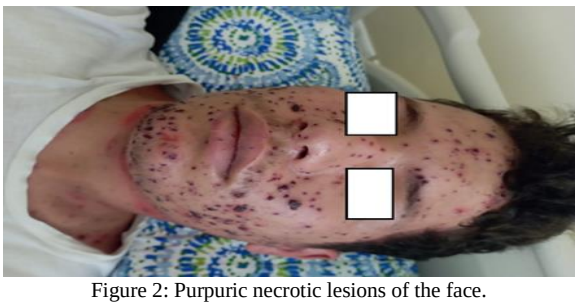

Figure 2: Purpuric necrotic lesions of the face.

III. DISCUSSION

Varicella is a ubiquitous infectious eruptive disease caused by a virus of the herpes family: varicella zoster virus (VZV), which is also responsible for shingles. The disease mainly affects children under 14 years of age [4;5]. Severe forms of the disease are frequently observed in immunocompromised patients due to the progress of immunosuppressive therapies, organ transplants and HIV infection [6;7].

Malignant varicella in immunocompetent adults is a

R. Bouagaga, H. Bennasser Alaoui, H. Bachir, S. Hamaz, and K. Serraj, "Malignant Varicella in an Immunocompetent Patient: A New Case Report," *International Research Journal of Pharmacy and Medical Sciences (IRJPMS)*, Volume 6, Issue 4, pp. 17-18, 2023.

serious form that can be life-threatening due to its multivisceral complications, particularly pulmonary and neurological, but also hematological, myocardial, etc… [5].

Varicella pneumonitis (PV) is the most frequent and serious complication, and reflects dissemination of VZV [1]. Its incidence is estimated to be between 16 and 33% and mortality can reach 20 to 50% [8; 9]. It occurs 1 to 7 days after the appearance of cutaneous signs [3]. The functional respiratory signs present in our patient (dyspnea, cough, and desaturation) point to pulmonary involvement, other signs such as pleuritic chest pain and hemoptysis are described in the literature [1]. However, PV can be discovered routinely on chest radiography [3]. Several risk factors for pulmonary involvement in varicella have been identified: male sex, adult age, smoking, number of skin lesions (>100 elements), pregnancy, close contact with an infected individual, any immunosuppression including leukemia, malignant lymphoma, pregnancy, systemic diseases, chronic respiratory diseases and HIV infection [1;3].

Our patient had risk factors such as age, male sex, skin involvement, and close contact with his two infected brothers. There is no gold standard for the diagnosis of varicella pneumonitis; the diagnosis can be made when there is a combination of pulmonary involvement in a high-risk area with concordant radiological features and a skin rash suggestive of varicella, in the absence of evidence of another cause [10]. Thoracic imaging often reveals more or less well-limited, diffuse nodules, reaching both lung fields, rarely unsystematized hilifugal opacities or heterogeneous infiltrates [1]. Bronchial endoscopy may demonstrate proximal vesicular endobronchial lesions [7]. Microbiological tests (viral culture, gene amplification methods) as well as biopsy and serology are not necessary when the clinical-radiological picture is typical [1].

Although PV remains the usual complication of malignant varicella in adults, other rare complications have been reported in the literature: neurological damage, which comes in second place, dominated by cerebellitis and meningoencephalitis [5;11], suspected in the presence of purpura fulminans and neurological disorders in a high-risk area; and haematological complications, the most common of which is post-infectious thrombocytopenia, which usually resolves in a few days [4].

The recommended curative treatment for malignant varicella is antiviral, preferably aciclovir, intravenously (10 mg/kg every eight hours), but also valaciclovir or ganciclovir for seven to ten days [1;7]. In addition to antiviral treatment, our patient received oxygen therapy and antibiotic therapy in response to signs of skin superinfection. The clinical and radiological evolution is often rapidly favourable due to the early management. However, cases of death in a situation of multivisceral failure have been reported in the literature [1].

IV. Conclusion

Varicella in healthy adults is rare, but often complicated by multivisceral attacks (pulmonary, neurological, haematological, hepatic, myocardial, etc.), which sometimes makes the diagnosis difficult; hence the importance of the clinic, which remains paramount (careful history and dermatological examination).

Conflicts of interest

The authors declare no conflicts of interest.

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