Journal of african clinical cases and reviews / Journal africain des cas cliniques et revues

www.jaccrafrica.com ISSN 2712-6412 Open access

Case report

Juvenile melanoma of the buttock in a 15-year-old black girl

Mélanome juvénile de la fesse chez une adolescente noire de 15 ans

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Résumé

Introduction : Le mélanome juvénile est une tumeur maligne rare développée aux dépens des mélanocytes chez les sujets de moins de 19 ans. Il représente 1 à 3 % des cancers pédiatriques et survient préférentiellement au niveau du tronc et des extrémités. Son diagnostic est souvent retardé en raison d'une présentation clinique atypique, particulièrement chez les patients à peau foncée, ce qui compromet le pronostic. Nous rapportons un cas de mélanome de la fesse chez une adolescente de 15 ans à phototype VI.

Rapport de cas : Une adolescente de 15 ans, de phototype VI, sans antécédents particuliers, consultait pour une tuméfaction de la fesse gauche évoluant depuis un an. L'examen retrouvait une masse sous-fessière arrondie, lisse, vascularisée, mesurant 5 cm. Une exérèse simple a été réalisée. L'étude anatomopathologique a mis en évidence une prolifération maligne de mélanocytes atypiques infiltrant le derme réticulaire, avec une épaisseur de Breslow de 15 mm et un niveau de Clark V. L'immunohistochimie montrait une positivité des cellules tumorales pour la protéine S100, confirmant le diagnostic de mélanome. Aucun bilan d'extension ni biopsie du ganglion sentinelle n'ont été réalisés en raison de l'absence de suspicion diagnostique initiale et la patiente a été perdue de vue après l'intervention.

Discussion : Le mélanome pédiatrique est exceptionnel, avec une incidence estimée à 5–6 cas par million d'enfants par an. Il touche principalement les populations caucasiennes et reste

rare chez les sujets à phototype VI, chez lesquels il siège habituellement aux extrémités acrales. Chez l'enfant, les critères cliniques classiques ABCDE sont souvent absents, les lésions pouvant être amélanotiques, nodulaires ou d'aspect bénin, expliquant les retards diagnostiques. Le diagnostic repose sur l'examen histologique, souvent complété par l'immunohistochimie (S100, Melan-A, HMB45). Le pronostic dépend essentiellement de l'épaisseur de Breslow, du niveau de Clark, de l'ulcération et de l'atteinte ganglionnaire. Dans notre observation, l'absence de marges de sécurité, l'absence de bilan d'extension et de biopsie du ganglion sentinelle, liées aux contraintes techniques et à la perte de vue de la patiente, constituent des facteurs péjoratifs. La prise en charge optimale repose sur une exérèse large, l'évaluation ganglionnaire et, selon le stade, des traitements adjuvants (immunothérapie, thérapies ciblées) associés à une surveillance prolongée.

Conclusion : Le mélanome juvénile est une affection rare dont le diagnostic est particulièrement difficile chez les sujets à phototype foncé en raison de présentations cliniques atypiques. Cette observation illustre les défis diagnostiques et thérapeutiques rencontrés en milieu à ressources limitées. Une suspicion clinique précoce, une confirmation histopathologique avec immunohistochimie et une prise en charge conforme aux recommandations sont essentielles pour améliorer le pronostic.

Mots-clés : Mélanome juvénile ; Mélanome pédiatrique ; Phototype VI ; Immunohistochimie ; Pays à ressources limitées.

Abstract

Introduction: Juvenile melanoma is a rare malignant tumor arising from melanocytes in individuals under 19 years of age. It accounts for 1–3% of pediatric cancers and most commonly affects the trunk and extremities. Diagnosis is frequently delayed because of its atypical clinical presentation, particularly in patients with dark skin, resulting in a poorer prognosis. We report a case of buttock melanoma in a 15-year-old girl with phototype VI.

Case Report: A 15-year-old girl with phototype VI and no significant medical history presented with a one-year history of a progressively enlarging left buttock mass. Clinical examination revealed a smooth, rounded, vascularized subgluteal mass measuring 5 cm. Simple surgical excision was performed. Histopathological examination demonstrated a malignant proliferation of atypical melanocytes infiltrating the reticular dermis, with a Breslow thickness of 15 mm and Clark level V. Immunohistochemistry showed diffuse S100 positivity, confirming the diagnosis of melanoma. No staging investigations or sentinel lymph node biopsy were performed because melanoma was not suspected preoperatively, and the patient was subsequently lost to follow-up.

Discussion: Pediatric melanoma is an exceptionally rare malignancy, with an estimated incidence of 5–6 cases per million children annually. It predominantly affects Caucasian populations and is uncommon in individuals with phototype VI, in whom acral sites are the usual locations. In children, the classic ABCDE criteria are often absent, as lesions may be amelanotic, nodular, or mimic benign tumors, contributing to delayed diagnosis. Histopathological examination, supported by immunohistochemistry, remains the cornerstone of diagnosis. Prognosis depends mainly on Breslow thickness, Clark level, ulceration, and lymph node involvement. In the present case, the absence of adequate surgical margins, staging investigations, and sentinel lymph node biopsy—due to limited resources and loss to follow-up—represented major adverse prognostic factors. Optimal management includes wide local excision, lymph node assessment, stage-adapted adjuvant therapy, and long-term surveillance.

Conclusion: Juvenile melanoma is a rare malignancy whose diagnosis is particularly challenging in patients with dark skin because of its atypical clinical presentation. This case highlights the diagnostic and therapeutic challenges encountered in resource-limited settings. Early clinical suspicion, histopathological confirmation with immunohistochemistry, and guideline-based management are essential to improve patient outcomes.

Keywords: Juvenile melanoma; Pediatric melanoma; Phototype VI; Immunohistochemistry; Resource-limited settings.

Introduction

Juvenile or pediatric melanoma is a malignant tumor proliferation arising from melanocytes and occurring in children under 19 years of age [1]. It is rare, even exceptional, representing 1 to 3% of all childhood malignancies [2] and 5% of skin tumors [3]. The extremities and trunk are the preferred locations for juvenile melanoma [2]. It can develop on healthy skin (de novo) but most often in the context of a specific condition (mole, etc.) [4]. Its prognosis remains poor due to its atypical clinical presentation, which delays treatment.

We report a case of melanoma of the left buttock in a 15-year-old girl.

Case report

This is a 15-year-old adolescent with phototype VI and no significant past medical history. She consulted for a swelling of the left buttock that had been developing for one year. Clinical examination revealed a good general condition and a regular, smooth, rounded, and vascular mass measuring 5 cm along its longest axis in the right subbuttock. The mass was excised and sent for histopathological examination. Macroscopic examination revealed a firm mass, partially covered by a skin flap, weighing 40 g and measuring 7 x 5 x 3.5 cm. It was whitish-gray with heterochromic, whitish-brown tissue sections on cross-section (Figure 1). Histology showed a malignant tumor proliferation composed of atypical melanocytes, elongated, sometimes round, and highly atypical, focally producing melanin pigments (Figure 2). The tumor infiltrated the reticular dermis without a safety margin, with a Breslow thickness of 1.5 cm and a Clark level V. Immunohistochemical analysis showed nuclear and cytoplasmic staining of the tumor cells with S100 (Figure 3). As the clinical presentation was not suggestive of melanoma preoperatively, no pre-treatment imaging was performed. The patient was lost to follow-up after the procedure.

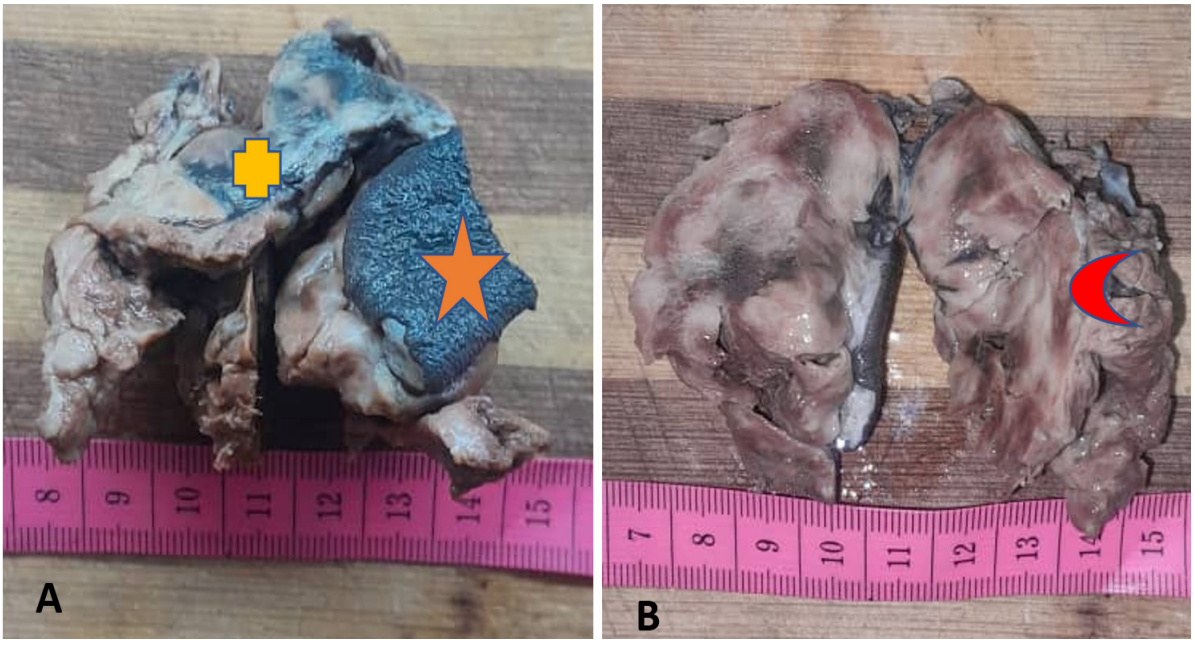


Figure 1: A: Brownish tissue mass partially covered by a skin flap (orange star) with necrotic changes (yellow cross). B: Brownish section (red crescent).

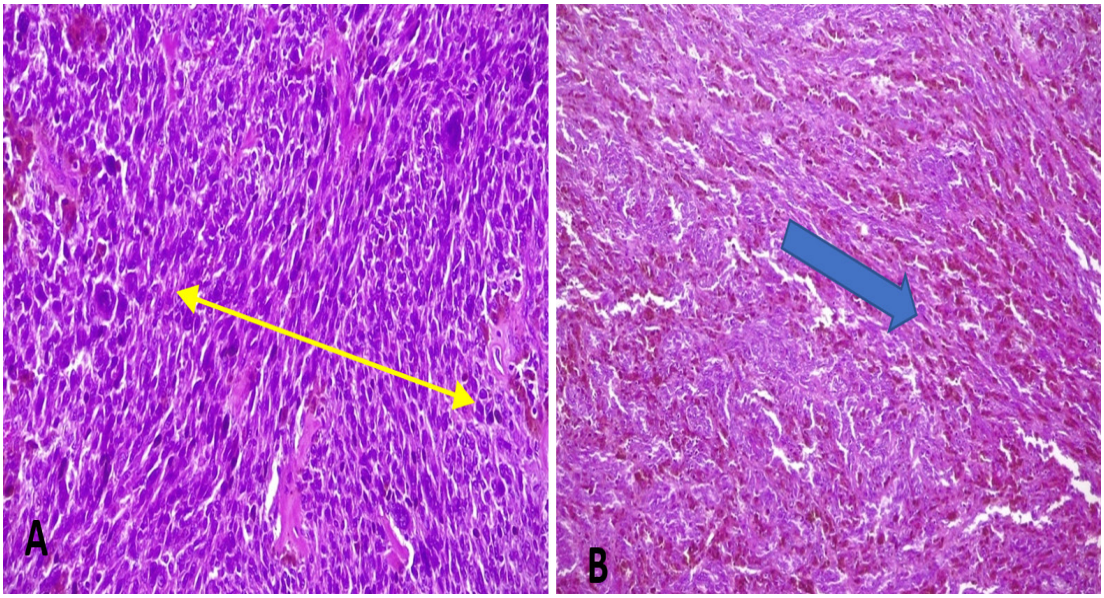


Figure 2: A: Malignant tumor proliferation composed of large, elongated, sometimes round, highly atypical cells (double-headed arrow). B: Tumor cells producing melanin pigments (blue arrows).

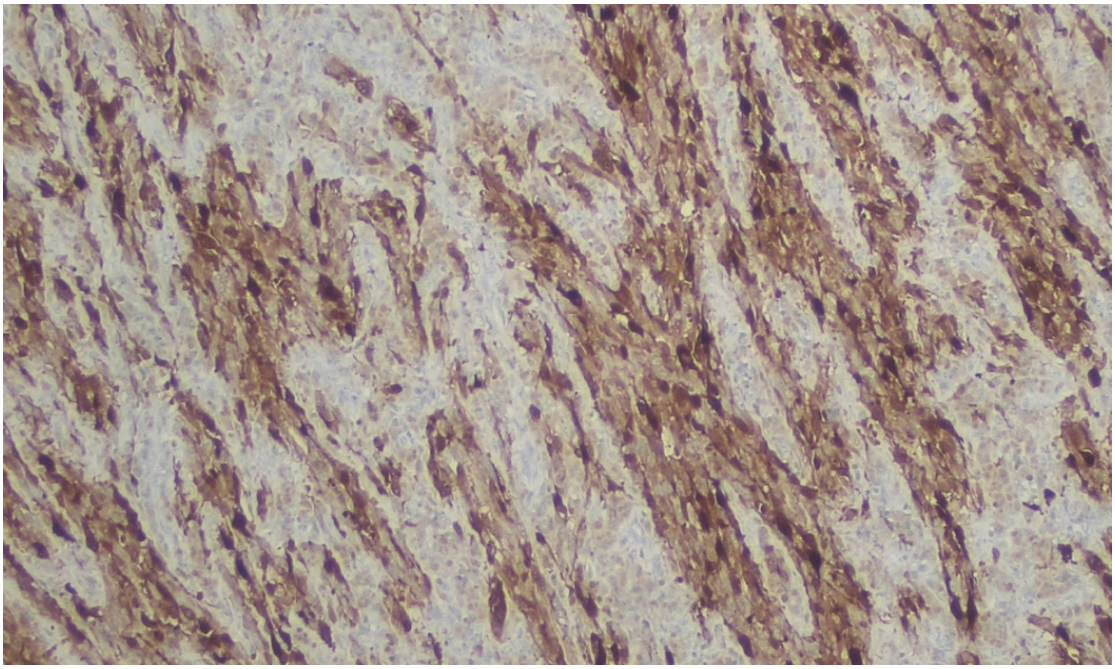


Figure 3: Diffuse Cytoplasmic and nuclear positivity \of tumor cells for S100 IHC x 150 ACP UCAD.

Discussion

Melanoma is the deadliest form of skin cancer, and its incidence is increasing worldwide. The incidence of melanoma is approximately 5 to 6 cases per million in children under 20 years of age [5]. It occurs almost exclusively in white populations, while its incidence remains very low in populations of African or Asian descent. The highest incidence rates are recorded in Australia and the southern United States [6]. Pediatric melanoma encompasses a fairly heterogeneous group of patients, including newborns, children, and adolescents, with a variety of distinct clinical presentations. Childhood melanoma can be broadly subdivided into two categories: diseases presenting in early childhood (0-9 years) and those presenting in middle childhood and adolescence (10-17 years). This cancer is exceptional in children, representing 1 to 3% of pediatric malignancies [1], and 0.7% of all melanoma cases [7]. The absolute numbers

remain low, but its incidence has increased by 50% in patients under 20 years of age over a 20-year study period. The reasons for this increase are unclear and are probably multifactorial (congenital melanocytic nevi, prolonged sun exposure and sunburn, indoor tanning, genetic factors, xeroderma pigmentosum, family history of melanoma) [8]. No family history of melanoma was found in our patient. The sex ratio is 1:1 but varies with age. There is a slight male predominance between one and four years of age, a trend that reverses over time (girls represent 39.3% of melanomas between one and four years of age and 60% between ten and 19 years of age) [4]. In individuals with phototype VI, melanoma is very rare, accounting for less than 2% of skin cancers [2]. It preferentially affects acral areas: palms, soles, and nail folds, as well as mucous membranes. When faced with a progressive pigmented lesion in a child, the differential diagnosis should primarily include a congenital nevus, a birthmark, or post-inflammatory hyperpigmentation. Diagnosis is often delayed in individuals with dark skin because dark pigmentation masks the classic A-B-C-D-E skin type criteria. Furthermore, low clinical suspicion delays diagnosis and contributes to a more guarded prognosis.

Several reports have suggested that juvenile melanoma presents distinct biological, clinical, and histopathological characteristics compared to adult melanoma [2].

Adolescent melanomas are most often located on the trunk and extremities [2]. As with adult melanoma, there is a tendency for males to present on the trunk, while females more frequently affect the lower extremities. This is the case for our patient, a 15-year-old girl with an unusual location in the left buttock.

The classic ABCDE characteristics should raise suspicion of melanoma: [9] A=Asymmetry, B=Irregular, notched, polycyclic, or well-defined border, C=Multiple colors or uneven distribution of colors (mixed color), D=Diameter > 6 mm, E=Evolution, i.e., the tendency to change rapidly over weeks or months.

Our patient's tumor is rounded, smooth, and vascular, measuring 5 cm. However, childhood melanomas, particularly in prepubertal cases, deviate from the ABCDE criteria [10]: pink or reddish, dome-shaped papule, amelanotic and nodular lesion, often mimicking benign lesions (symmetrical, pigmented lesions with regular borders). Modified ABCD criteria: amelanotic, bleeding/bump, uniformity of color, de novo/any diameter [10].

The prognosis for juvenile melanomas is often poor, representing 0.1% of all melanoma deaths recorded in the SEER (Surveillance, Epidemiology, and End Results) database [7]. It depends on Breslow thickness, Clark level, presence or absence of ulceration, diagnosis in adolescence, and location on the head and neck. Our patient had a high Clark level (V), a Breslow thickness >4 mm, and no ulceration. Given this, the patient's prognosis is guarded.

The formal diagnosis is made histologically. However, the histology of melanoma in children is difficult to establish, as demonstrated by numerous retrospective studies in which a significant number of melanomas were reclassified after examination as nevi [11]. Conversely, there are also documented melanomas that have led to distant metastases and the patient's death, and which presented the histological characteristics of a Spitz nevus [12]. These tumors are located deep within the dermis and hypodermis and contain epithelioid or spindle cells, and sometimes small round cells mimicking an infantile round cell tumor, making diagnosis challenging. Mitoses, areas of necrosis, ulceration, and signs of inflammation are generally present.

Immunohistochemistry showed staining of tumor cells by Melan A, S100, HMB45, and a high Ki-67. Only S100 was used in our patient and was positive.

Juvenile melanomas can be confused with other melanocytic tumors such as benign nevi, Spitz nevi, bruises, and pyogenic granulomas. These will be quickly ruled out by histology.

The first-line treatment is wide local excision of the deep fascia. The recommended margins in adults are 1 to 2 cm depending on the Breslow thickness. In children, they are not well defined but, in most

cases, follow the guidelines for adult melanoma [13]. Sentinel lymph node biopsy (SLNB) should be individualized, taking into account factors such as the patient's age, clinical circumstances, melanoma subtype, diagnostic certainty, location on the body, lymphatic drainage pattern, and desired prognostic information [13]. Consider SLNB if the lymph node is < 0.8 mm thick with ulceration or 0.8 to 1.0 mm thick with or without ulceration [13]. For melanomas with a Breslow thickness > 1 mm, international guidelines recommend sentinel lymph node biopsy for staging. This procedure helps identify patients at high risk of metastasis and guide the indications for adjuvant therapies. In our case, SLNB was not performed. This omission is explained by the unavailability of this technique at our institution and by the patient's loss to follow-up. A clinical examination and ultrasound of the draining lymph node areas could have been offered as an alternative. This limitation highlights the inequalities in access to standard care in resource-limited countries and the impact on patient prognosis. In our patient, the mass excision was incomplete without a safety margin due to the absence of preoperative diagnostic suspicion. Preoperative clinical images were not available. Furthermore, no lymph node dissection was performed. Given her Clark and Breslow thicknesses, her prognosis is not only guarded, but there is also a major risk of recurrence and secondary localization.

Adjuvant targeted therapy is also necessary, including the use of BRAF and MEK inhibitors, immunotherapy with checkpoint inhibitors, PD-1 and CTLA-4 inhibitors, chemotherapy, and radiotherapy [14]. Finally, surveillance with regular examinations of the entire body skin and lymph nodes is essential [13].

As the patient was lost to follow-up, surgical revision and staging could not be performed. These limitations reflect the difficulties in managing the condition in our resource-limited setting.

Conclusion

Juvenile melanoma is a rare entity, especially in individuals with phototype VI. This case illustrates the diagnostic and therapeutic challenges in our resource-limited setting, where the often atypical clinical presentation and low suspicion delay diagnosis and compromise the prognosis. Histology is particularly challenging, frequently requiring referral to expert centers. Optimal management relies on wide excision from the outset and assessment of lymph node status. This case underscores the need for increased vigilance in the presence of any evolving pigmented lesion in children, regardless of phototype, in order to improve the prognosis in our region.

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Available online : July 31, 2026

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Conflict of interest : None

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To cite this article

K Dansokho, M Wade, PI Dione, AM Gaye, GNC Deguenonvo, MJ Dieme-Ahouidi et al. Juvenile melanoma of the buttock in a 15-year-old black girl. Jaccr Case Reports 2026; 1(2): 1-7

https://doi.org/10.70065/2612.jaccrCasereports.001L013107