The Spitz Nevus: Review and Update

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KEYWORDS

• Spitz nevus • Atypical Spitz nevus
• Spindle and epithelioid cell nevus • Skin neoplasm
• Borderline melanoma • Spitz tumor
• Childhood melanoma • Juvenile melanoma

The Spitz nevus is a melanocytic lesion that has melanoma-like features on light microscopic examination but in typical form is distinguishable from melanoma, and usually exhibits benign behavior. There are also Spitz-like melanomas with microscopic features of Spitz lesions but otherwise distinctly malignant microscopy. Finally, borderline Spitz lesions have features of Spitz lesions and features of melanoma, and are recognized as of uncertain biologic potential and possible melanoma. These more rare borderline lesions are the most controversial regarding diagnosis and treatment. However, even Spitz nevi with typical histology have on very rare occasions unexpectedly led to metastasis and death.[1–3] Thus, even though Spitz lesions are easily recognized as distinct, they can defy current light microscopic diagnosis principles that are traditionally used to distinguish those malignant versus nonmalignant. There are many unanswered questions about the biologic nature of Spitz nevi. The diagnosis, prognosis, management, and even nomenclature of these lesions is controversial.

Despite improvements in diagnosis and combined experience over the past century since the Spitz lesion was first described,[4] this area remains one of the most controversial in pathology. To date investigations searching to more accurately diagnose and effectively treat these lesions have included limited numbers of cases and suffer from cohort bias. Multiple diagnostic and management strategies have been proposed and are individually utilized. Recently described investigational techniques seem promising. At this point, multicenter studies are needed to collectively determine whether they can be relied on to more accurately predict prognosis and therefore improve management.

EPIDEMIOLOGY

Spitz nevi are common in children but relatively uncommon in adults. Spitz nevi account for approximately 1% of excised nevi in children.[5] Many different names have been used for Spitz nevi since the original description of juvenile melanoma (**Box 1**), in an attempt to assign probable behavior. The names reflect a spectrum from benign to malignant, based on light microscopic analysis. Melanoma is probably best reserved in the name for Spitz lesions when referring to frank melanoma that has Spitz features (“spitzoid melanoma”) or in Spitz lesions with features of Spitz and melanoma where biologic behavior is not predictable (“borderline melanoma”). The term “nevus” connotes benign behavior and is probably best reserved for lesions on the benign end of the spectrum or when atypical features are present if used with a qualifying description (“Spitz nevus with atypical features”). The term lesion is more neutral and can more broadly encompass all “Spitz lesions.” Urso[6] and Piepkorn[7] suggest use of the term Spitz “tumor” to distinguish all Spitz lesions as separate entities from nevus and

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Box 1
Spitz nevus synonyms

Spitz nevus synonyms
- Juvenile melanoma
- Spindled and epithelioid cell nevus
- Pigmented spindle cell nevus of Reed
- Nevus of large spindleoid or epithelioid cells

Synonyms for Spitz that are neutral
- Spitz lesion
- Spitz tumor
- Spitzoid lesion; spitzoid tumor; spitzoid melanocytoma; spitzoid neoplasm

Atypical or borderline Spitz synonyms
- Atypical Spitz nevus
- Melanocytic tumor of uncertain potential (MELTUMP); melanocytic lesion of uncertain malignant potential (MUMP); Spitz tumor if uncertain potential (STUMP)
- Borderline melanoma

melanoma therby emphasizing separate diagnostic and biologic paths.[6,8] Individual names are used inconsistently between various investigators, and thus attention should be paid to the individual author's application of the terminology.

HISTORY

The Spitz nevus has been recognized in the literature since the early twentieth century, when Darier and Civatte[4] described a fast-growing red nodule on the cheek of a young boy and reported that they were unable to decide by light microscopy whether it was a melanoma or not. The pathology of the lesion they described, which would later be termed a Spitz nevus, was indistinguishable in their viewpoint from melanoma. Similar lesions would normally have been reported as malignant melanoma at this time; however, some researchers such as Darier and Civatte were beginning to recognize that these growths could exhibit benign clinical behavior. Still, the only criterion for distinguishing these lesions from melanoma was age of the patient. In 1948, Sophie Spitz[9] described in detail the light microscopic features of a group of these lesions, which she termed juvenile melanoma, and the diagnosis became widely recognized as distinct. Spitz compared the pathology of juvenile melanoma in individuals 18 months to 12 years old to melanoma in children 14 to 19 years old and to melanocytic nevi in children. She described common features of these juvenile melanoma lesions, and emphasized the presence of giant cells as a distinguishing feature from postpubertal melanoma.

Spitz believed that these juvenile melanomas were unable to metastasize until adulthood because of a hormonal effect. The indicated treatment for juvenile melanoma in these years was removal before adulthood to prevent malignant degeneration. There were 2 problems with this assumption that she would later recognize: (1) a 12-year-old girl included in the original Spitz description died of metastasis,[2] and (2) benign Spitz lesions were found in adults as well. Both these factors implied that age was not the only factor in determining benign verses malignant behavior.

Allen and Spitz[10] later described a refined description of pathologic criteria for distinguishing Spitz nevi from melanoma in 1953. Arthur Allen, Sophie Spitz's husband,[11] eventually reviewed the pathology of the 12-year-old in the original description who died from melanoma and determined that the pathology was, in fact, not consistent with the diagnosis of juvenile melanoma.[2] Thus, the difficulty in pathologic diagnosis of Spitz nevi was apparent as early as the first description. Although more refined criteria for diagnosis of Spitz nevus are available today, the biologic potential remains unpredictable in a subset of lesions with atypical features that resemble melanoma.

CLINICAL PRESENTATION

Spitz nevi occur most often in children or young adults[12] but can occur at any age. Nearly half to two-thirds of Spitz nevi occur in individuals younger than 20 years.[13] Spitz nevi become less common with increasing age, and they are more likely to be diagnosed as melanoma with increasing age of the patient. Congenital Spitz nevi have been reported.[14,15] The lesions are more common in Caucasians with fair skin type and are slightly more frequently found in females.[5,16]

Spitz nevi commonly present on the face, head, neck, or lower extremities, but can occur anywhere on the body. In one study, Spitz nevi in children were more commonly on the head and neck whereas Spitz nevi in adults were more commonly located on the extremities (consistent with the observation in children that melanocytic nevi are found in greater frequency on the head and neck). The lesions are frequently solitary, but multiple and agminated (multiple grouped) lesions can occur. Grouped lesions may coalesce on