Introduction

Juvenile papillomatosis of the breast ("Swiss cheese disease") is a benign localized proliferative condition first described by Rosen and colleagues in 1980 (5). Patients with this lesion often have a family history of breast carcinoma, but rarely carcinoma may coexist with the lesion at the time of diagnosis. We present a case of a young male with juvenile papillomatosis of the breast. The pathology and clinical management of this rare lesion is discussed.

Case report

A 17 years-old male presented with 2-3 month history of intermittent bloody discharge from the right nipple and a slowly growing mass in the upper outer quadrant of the right breast. This mass measured approximately 2/3 cm, and was firm and well circumscribed. Microscopically, this lesion consists of ductal papillomatosis, apocrine and nonapocrine cysts, papillary apocrine hyperplasia, and duct stasis. This lesion occurs almost exclusively in women younger than 30 years of age. By review of the literature, we have only noted four cases of juvenile papillomatosis in male children, all of whom over age 11 years. We present a case of young male with juvenile papillomatosis of the breast, which represents uncommon description of papillomatosis lesions in either young infants or in young males.
Pathologic findings

The breast mass measured 1.5 × 0.6 × 0.4 cm. The external surface was yellow-tan. Bisection revealed multiple small cysts containing serosanguineous fluid. Microscopic sections showed dilated ducts with occasional hyperplastic epithelium, apocrine metaplasia, and foamy macrophages. Intraductal hemosiderin-laden macrophages were also noted. Structurally there was ductal ectasia with epithelial hyperplasia and hemorrhage, consistent with a diagnosis of juvenile papillomatosis. The cyst lining stained positive for cytokeratin and epithelial membrane antigen by immunostains.

Discussion

Juvenile papillomatosis of the breast is a benign localized proliferative condition that generally occurs in women younger over age 30 years. This disease was first described by Rosen in 1980 (5), and since that time a number of papers have been published which describe the particular pathologic and epidemiologic characteristics of this unique disease.

In young females, juvenile papillomatosis has been demonstrated to be a marker for the increased risk of breast cancer in the patient’s immediate family (7). The significance of a positive family history of breast cancer among first degree relatives of a patient with a borderline or “moderate risk” lesion of atypical or lobular hyperplasia is well established (7). In 1990, Rosen and Kimmel further assessed the long-term risk for a patient with juvenile papillomatosis, and found that patients with a positive family history for breast cancer and recurrent bilateral papillomatosis may be at increased risk for breast cancer themselves (6). In contrast to papillomatosis breast lesions in young women, juvenile papillomatosis of the male breast is an exceedingly rare condition. By extensive review of the literature, we found only four cases to date with juvenile papillomatosis of the breast. This presentation of a breast epithelial proliferative disorder in a young male represent a novel and previously undescribed clinical variant of juvenile papillomatosis. The effect of this condition on this patient’s risk of subsequent breast tumor development is unclear, as is the risk to this patient’s family for the development of breast cancer. We recommend that this patient and all other male or female children with juvenile papillomatosis should be followed clinically to detect further lesions which may require surgery.

Our case of young male represent one of reported cases to date with juvenile papillomatosis of the breast. This presentation of a breast epithelial proliferative disorder in a young male represent a novel and previously undescribed clinical variant of juvenile papillomatosis. The effect of this condition on this patient’s risk of subsequent breast tumor development is unclear, as is the risk to this patient’s family for the development of breast cancer. We recommend that this patient and all other male or female children with juvenile papillomatosis should be followed clinically to detect further lesions which may require surgery.

References