

Benign Umbilical Tumors Resembling Sister Mary Joseph Nodule

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ABSTRACT

BACKGROUND: When physicians see an umbilical nodule, most of them instinctively recall the Sister Mary Joseph nodule. Therefore, dermatologists need to recognize umbilical dermatoses that can be mistaken for the Sister Mary Joseph nodules. This study aimed to describe the different kinds of benign umbilical tumors as well as elucidate the factors that can be used to distinguish the Sister Mary Joseph nodule from these tumors.

METHODS: The “benign umbilical tumor” group included 19 patients, whereas the “Sister Mary Joseph nodule” group comprised 30 patients (2 from our department, 28 from PubMed search). We compared the clinical and dermoscopic findings between 2 groups.

RESULTS: In the “benign umbilical tumor” group, the most common diagnosis was dermatofibroma (5/19), followed by keloid (3/19), and soft fibroma (3/19). These tumors had various colors (red, brown to black, and flesh colored) and exhibit characteristic surface changes (eg, verrucous changes in epidermal nevi and verrucae). Conversely, most Sister Mary Joseph nodules have an erythematous color, oozing or ulceration on the surface, and nearby satellite lesions. Furthermore, the dermoscopic findings of Sister Mary Joseph nodules showed a polymorphous vascular pattern and a white or milky-red, amorphous area. Benign lesions showed different dermoscopic patterns: pigment networks with white areas (dermatofibromas), thrombosed capillaries (verrucae), and the “pore sign” (epidermal cysts).

CONCLUSIONS: Various cutaneous tumors can be mistaken for the Sister Mary Joseph nodule when they develop on the umbilicus; the clinical and dermoscopic differences found in this study may be useful for establishing a differential diagnosis.

KEYWORDS: Benign, dermoscopy, Sister Mary Joseph nodule, tumor, umbilicus

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Introduction

The umbilicus comprises the remnants of the umbilical cord and its adnexa, which are only present prenatally. It is composed of a cushion, which has a slightly declined border, and a cicatrix with an irregular base.¹ Umbilical metastasis from various types of cancer in the internal organs occurs because the umbilicus is located adjacent to the anterior peritoneum and because it communicates with many solid internal organs through a myriad of vascular and lymphatic drainage pathways.^{2–4} However, dermatoses of the umbilicus not only occur as a result of cutaneous metastasis, but can also be caused by inflammation, infection, or primary neoplastic conditions.

When physicians see an umbilical nodule, most of them instinctively recall the Sister Mary Joseph nodule. A Sister Mary Joseph nodule refers to a malignant umbilical tumor commonly associated with the metastases of internal solid organ cancer. This term was first described by Sister Mary Joseph in 1928.⁵ In this regard, dermatologists need to recognize other

umbilical tumors that can be mistaken for the Sister Mary Joseph nodule. In this study, we aimed to present the different kinds of benign umbilical tumors as well as elucidate the factors that can be used to distinguish the Sister Mary Joseph nodule from these tumors.

Materials and Methods

Study population and design

This study was retrospectively conducted from May 2007 to November 2016. A total of 21 patients with umbilical tumors who had visited one of the skin clinics of Pusan National University Hospital (Busan or Yangsan) were included in the study. The “benign umbilical tumor” group comprised 19 patients. The “Sister Mary Joseph nodule” group consisted of 30 patients: 2 from our department and 28 from a PubMed search using the keyword “Sister Mary Joseph nodule [title].”^{4–25}

The patients’ clinical photographs and dermoscopic findings were assessed, and their final diagnoses were confirmed based on the pathological reports. The digital dermoscopic images of the

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Table 1. Kinds of benign umbilical tumors reported in this study.

DIAGNOSIS	NO. (%)
Dermatofibroma	5 (26.3)
Epidermal cyst	1 (5.3)
Epidermal nevus	1 (5.3)
Hypertrophic scar	2 (10.5)
Keloid	3 (15.8)
Neurofibroma	1 (5.3)
Soft fibroma	3 (15.8)
Verruca vulgaris	3 (15.8)
Total	19 (100)

selected lesions were acquired using a dermoscope (DermLite II Pro HR equipment; 3Gen LLC., Dana Point, CA, USA; $\times 10$ magnification) attached to a digital camera (Sony Cyber-shot DSC-W290; Sony Corporation, Tokyo, Japan; $\times 5$ optical zoom, 12.1 megapixels).

We investigated the different kinds of benign umbilical tumors that occurred as well as the clinical and dermoscopic differences between benign umbilical tumors and the Sister Mary Joseph nodule. During the clinical assessment, the color, surface changes, shape, and consistency of the tumors were evaluated. In the PubMed-sourced cases of Sister Mary Joseph nodule, this assessment was made based on written descriptions as well as clinical images. However, the dermoscopic findings of the Sister Mary Joseph nodule were only described in 3 cases (2 from our own department and 1 from among the PubMed-sourced cases).

This study was approved by the Institutional Review Board (IRB) of Pusan National University Hospital (IRB number: H-1702-027-052), who waived the requirement for informed consent as this study was a retrospective study through a chart review.

Results

Kinds of benign umbilical tumors

More than 19 patients developed various kinds of benign umbilical tumors, which can be classified as epidermal and dermal neoplasms. Those of epidermal origin included epidermal cysts, epidermal nevi, and verrucae vulgaris, whereas those of dermal origin comprised dermatofibromas, hypertrophic scars, keloids, neurofibromas, and soft fibromas. Overall, dermatofibroma was the most common neoplasm (5/19; 26.3%); followed by keloid, soft fibroma, and verruca vulgaris (3/19; 15.8% in each case); hypertrophic scar (2/19; 10.5%); and epidermal cyst, epidermal nevus, and neurofibroma (1/19; 5.3% in each case). The overall results are summarized in Table 1.

Kinds of Sister Mary Joseph nodules

The age of the patients with a Sister Mary Joseph nodule ranged from 22 to 89 years with a mean age of 60.8 years. Most of these patients were women (28/30; 93.3%). The ovary was the most common primary cancer site in this group (11/30; 36.7%), followed by the pancreas (6/30; 20.0%), uterus (6/30; 20.0%), colon (3/30; 10.0%), testicle (2/30; 6.7%), stomach (1/30; 3.3%), and vulva (1/30; 3.3%). In terms of the histopathology, adenocarcinoma was the most common type (23/30; 76.7%), followed by squamous cell carcinoma (4/30; 13.3%), adenosquamous carcinoma (1/30; 3.3%), mixed tumor (1/30; 3.3%), and undifferentiated carcinoma (1/30; 3.3%).

Clinical and dermoscopic differences between benign umbilical tumors and the Sister Mary Joseph nodule

In terms of color, benign tumors showed various colors, including blue (epidermal cysts), dusky red (keloids), black (epidermal nevi), and skin tone (soft fibromas). By contrast, most (20/30; 66.7%) of the Sister Mary Joseph nodules exhibited a red color. Less often, they displayed an erythematous hue, whereas some appeared black (2/30; 6.7%), blue (1/30; 3.3%), skin tone (2/30; 6.7%), violet (4/30; 13.3%), and white (1/30; 3.3%) in color.

Regarding surface changes, many of the benign tumorous lesions (e.g. seborrheic keratoses, verrucae, and epidermal nevi) demonstrated verrucous changes. Conversely, the Sister Mary Joseph nodule showed oozing (15/30; 50.0%) and ulceration (11/30; 36.7%) on the surface, and clinical examination revealed the formation of satellite lesions (26.7%; 8/30) in the vicinity of the umbilical nodule. Although none of the benign tumors had a fixed consistency, most of the Sister Mary Joseph nodules had a firm and fixed consistency (93.3%; 28/30).

On dermoscopy, benign umbilical tumors showed various patterns consistent with their histological nature: pigment network with a white area (dermatofibroma), thrombosed capillaries (verruca), diffuse pigmentation (epidermal nevus), and the "pore" sign (epidermal cyst). By contrast, the Sister Mary Joseph nodules exhibited a polymorphous vascular pattern and a white or milky-red amorphous area (Figure 1). The overall results are summarized in Table 2.

Discussion

The umbilicus is located at the center of the abdomen; hence, it can be easily evaluated during clinical examinations. Physicians consider the umbilicus as an important part in the diagnosis of certain conditions. The umbilicus itself constitutes the remnants of the umbilical cord and its adnexa, which are only present during the prenatal period.¹ However, any unusual changes that occur in the umbilicus are considered significant by physicians, as the greater part of the venous and lymphatic drainage of many solid internal organs passes through this area;

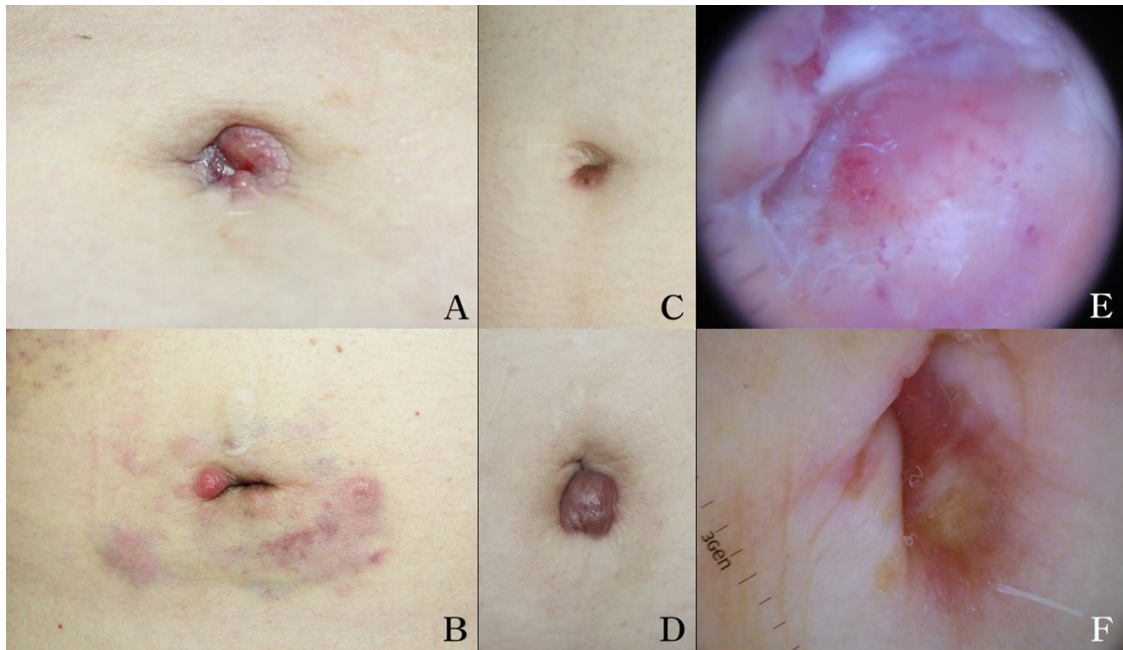


Figure 1. Umbilical tumors (clinico-dermoscopic images). (A, B) Clinical photograph of a Sister Mary Joseph nodule showing erythematous color and satellite lesion in the vicinity of the nodule; (C) clinical photograph of a dermatofibroma; (D) clinical photograph of a keloid; (E) dermoscopic finding of a Sister Mary Joseph nodule showing polymorphous vascular pattern and white, milky-red structureless area; (F) dermoscopic finding of a dermatofibroma showing a pigment network with white area.

Table 2. Clinical and dermoscopic differences between benign umbilical tumors and Sister Mary Joseph nodule.

	BENIGN UMBILICAL TUMORS (N=19)	SISTER MARY JOSEPH NODULE (N=30)
Color	Various color ^a	Various but dominantly red ^b
Satellite lesion	None	Some (26.7%)
Surface change	Verrucous in epidermal tumors	Oozing (50.0%) or ulcer (36.7%)
Consistency	Not fixed	Firm and fixed (93.3%)
Dermoscopic finding	Specific patterns corresponding to its nature ^c	Polymorphous vascular pattern and white or milky-red structureless area

^aBlue, epidermal cyst; dusky red, keloid; black, epidermal nevus; and skin tone, soft fibroma.

^bRed color (20/30, 66.7%). Besides erythematous hue, black (2/30, 6.7%), blue (1/30, 3.3%), skin tone (2/30, 6.7%), violet (4/30, 13.3%), and white (1/30, 3.3%).

^cPigment network with white area in dermatofibroma, thrombosed capillaries in verruca, diffuse pigmentation in epidermal nevus, and the “pore” sign in epidermal cyst.

the umbilicus communicates with the aforementioned organs through these vessels.^{2,3} Therefore, the umbilicus often serves as a passage for malignant tumors that originated from the internal solid organs. When malignancies of the internal organs spread, skin metastases in the umbilicus—whose primary cancers are usually found in the pelvic or abdominal region—are dubbed “Sister Mary Joseph nodules” after the surgical assistant Sister Mary Joseph Dempsey, who first suggested the concept of metastatic umbilical tumors to Dr. Mayo—founder of the Mayo clinic.^{5,10} Dr. Mayo subsequently studied these tumors and published an article about them. Gastrointestinal malignancies account for about half of the underlying pathologies, and men are more likely to have an underlying cancer of the gastrointestinal tract.⁷ Similarly, gynecological cancers account for about 1 in 4 cases.⁴ Appearance of the Sister Mary Joseph nodules is a metastatic sign of an internal malignancy

and can therefore indicate advanced primary disease and poorer prognosis.^{20,24}

Aside from these malignant conditions, various benign neoplasms can develop on the umbilicus,²⁶ namely, epidermal cysts, angiofibromas, keloids, endometrioses, nevi, teratomas, and papillomas.^{26–28} However, no study has investigated which benign tumors can occur on the umbilicus. In this study, we described various benign umbilical tumors, including dermatofibromas, soft fibromas, neurofibromas, epidermal nevi, and epidermal cysts. Of these, dermatofibromas, soft fibromas, and neurofibromas have not been reported as benign umbilical tumors in previous studies.

Among skin malignancies, dermatofibrosarcoma protuberans (DFSP) can be mistaken for a benign nodule, but its prevalence is not high. The clinical presentation of malignant nodule can be confused with that of nonmelanoma skin cancers such

as basal cell carcinoma, squamous cell carcinoma, and, in rare cases, Merkel cell carcinoma and DFSP.

Therefore, clinicians should be able to distinguish Sister Mary Joseph nodules from various benign umbilical tumors because differential diagnosis will help improve future evaluations and therapeutic approaches. In this regard, radiological imaging and histopathology are occasionally needed for differential diagnosis. However, these techniques have some shortcomings; for instance, the cost of performing an imaging examination is high, and histopathological evaluation is an invasive method. Therefore, it may be important to perform a differential diagnosis based on only the patients' clinical information.

In this regard, benign umbilical tumors exhibited various colors: blue (epidermal cysts), brown (keloids), black (epidermal nevi), and skin tone (soft fibromas). Conversely, most Sister Mary Joseph nodules had a red color, perhaps because they had an abundant vascular supply and because angiogenesis is uncontrolled in malignant tumors.²⁹ Regarding the physical findings, Sister Mary Joseph nodules showed oozing (15/30; 50.0%) and ulceration (11/30; 36.7%) on the surface, as well as satellite lesions (26.7%; 8/30) in the vicinity of the umbilical nodule. In benign umbilical nodules, the surface changes were not relatively variable, although verrucous changes were observed in seborrheic keratosis, verruca, and epidermal nevus. Oozing or ulcerative changes on the surface indicate that a malignant tumor is undergoing necrotic degeneration due to its rapid growth.⁶ Similarly, multiple, widely distributed satellite lesions reflect malignancy.²⁴

In this study, none of the benign umbilical tumors were fixed, whereas most of the Sister Mary Joseph nodules (93.3%; 28/30) showed a firm and fixed consistency upon physical examination, indicating packed cellularity and invasion into the adjacent tissues, both of which are characteristics of malignancy.²⁹

On a separate note, dermoscopy can provide information that can help in differentiating benign umbilical tumors from Sister Mary Joseph nodules.^{14,30-33} It is a noninvasive, quick, and easy diagnostic method that can be performed in the clinic. Furthermore, it allows researchers to visualize the anatomical structures that cannot be seen by the naked eye. Meanwhile, only 1 study reported the following dermoscopic findings of a Sister Mary Joseph nodule: a polymorphous vascular pattern, a milky-red area, and a white amorphous area.¹⁴ Nonetheless, these findings occurred in both our cases with Sister Mary Joseph nodule. The characteristic polymorphous vascular pattern and the white and milky-red area are considered as the pathognomonic signs of neoangiogenesis corresponding to a malignant growth or metastatic skin cancers.¹⁴ By contrast, various benign tumors showed different specific patterns, depending on their nature, such as pigment network with white area (dermatofibromas),³⁰ thrombosed capillaries (verrucae),³¹ diffuse pigmentation (epidermal nevi),³² and the "pore sign" (epidermal cysts).³³ Although the incidence of Sister Mary Joseph nodule is relatively low nowadays, the occurrence of symptoms indicates a serious

condition and should be managed as soon as possible. In this regard, the clinical and dermoscopic differences described in this study could be helpful to physicians treating patients with umbilical tumors. In the early stage of Sister Mary Joseph nodules, they appeared as a papule. Most of the Sister Mary Joseph nodules, or umbilical metastasis, appeared red in color, were hard, and had a fixed consistency. On dermoscopy, the Sister Mary Joseph nodules exhibited a polymorphous vascular pattern.

This study had some limitations. First, it was based on the assumption that most physicians instinctively recall the Sister Mary Joseph nodule when they see an umbilical nodule in a clinical context. Furthermore, the data regarding the Sister Mary Joseph nodules were mostly derived from a PubMed search. Finally, we intended to ascertain the dermoscopic characteristics of the Sister Mary Joseph nodule; however, only a few studies reporting these characteristics have been published. Therefore, future larger studies are warranted in the further to validate the findings of our study.

Conclusions

When physicians see an umbilical nodule in a clinical context, most of them instinctively recall the Sister Mary Joseph nodule. However, according to our present study, various benign tumors can develop on the umbilicus. The clinical and dermoscopic differences described in this study may help in differentially diagnosing benign neoplastic tumors and Sister Mary Joseph nodules.

Author's Note

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Author Contributions

M-B.K. contributed to study conception and design, and M-Y.Y., J-O.S., H-S.K. to data collection. B-S.K., M-B.K. contributed to data analysis, and D-L.H. to draft writing. D-L.H., M-B.K. contributed to revision, and all authors read and approved the final manuscript.

Ethics approval and consent

This study was approved by the Institutional Review Board (IRB) of Pusan National University Hospital (IRB number: H-1702-027-052), who waived the requirement for informed consent as this study was a retrospective study through a chart review.

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