Review Article

Adult onset pityriasis rubra pilaris

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ABSTRACT

Pityriasis rubra pilaris (PRP) has always been an intriguing topic ever since its inception. It is a group of chronic disorders characterized by reddish orange plaques with pityriasiform scaling showing follicular keratoses, palmoplantar keratoderma, and sometimes, erythroderma. It occurs all over the world but with racial variations. Its incidence might vary and the age at onset, behavior, clinical appearance, and prognosis are considered to be very important for its classification. It may manifest either as Type I classical adult onset PRP, Type II atypical adult (onset) PRP, or Type VI PRP (HIV-associated PRP pityriasis rubra pilaris) in contrast to classical juvenile (Type III) and circumscribed juvenile (Type IV) encountered among children. Its diagnosis is largely clinical with microscopic pathology being a useful supplement, but it continues to be a therapeutic dilemma. We review the epidemiology of adult onset PRP here and take stock of the prevalent treatment options.

Key Words: Adult onset, Pityriasis rubra pilaris

INTRODUCTION

Ever since the first reported case of the disease, pityriasis rubra pilaris (PRP) has remained a consistently recorded and researched entity to date.[1-12] However, its etiology and management have remained a challenge for the treating physician. It is seen in adults (adult onset) as well as in children, and affects both the sexes. Occasionally, PRP is associated with other diseases and it was speculated that the disorder might be the result of an abnormal immune response to some antigenic stimuli.[4] However, familial occurrence of the disease might point to some genes that predispose the individual to develop this disorder after certain precipitating events.[13-17] The occurrence of this dermatosis in association with human immunodeficiency virus (HIV)/acquired immunodeficiency disease (AIDS) patients has sparked a dialogue as to whether or not it is yet another variant of PRP.[18-22]

DEFINITION

Pityriasis rubra pilaris refers to a group of chronic disorders characterized by reddish orange plaques with pityriasiform scaling showing follicular keratoses, palmoplantar keratoderma, and sometimes, erythroderma. Familial as well as acquired forms of the disease have been reported.[23]

HISTORY

Devergie has been credited with the naming of 'pityriasis pilaris' in 1857, which received the eponym of Devergie's diseases.[24] However, much before that, Tarral[1] in 1835, recorded the case description of this disease in the "Rayer's[1]-a theoretical and practical treatise on the disease of the skin" under the title of 'general psoriasis'. Devergie stressed that Tarral's case was an example of PRP, and observed that PRP might be confused with psoriasis. However, in 1889, Besnier[25] advanced the present day name, "pityriasis rubra pilaris." Later, several authors[26-31] recognized that PRP was of many types, and thus suggested several working classifications.[32-37] With the advent of HIV/AIDS, its association with PRP has been noticed by many, prompting expansion of the existing classification to accommodate PRP associated with HIV as a distinct type.[20,38-40]

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EPIDEMIOLOGY

Although PRP occurs worldwide, there are racial variations.[2,3] Its incidence might vary-it is 1 in 5,000 in Great Britain[34] and 1 in 50,000 in India[31] in an outpatient setting. Both the sexes are affected equally at all ages.[14,33] A bimodal or trimodal age distribution has been recorded with peak incidence in the 1st, 2nd and 6th decade of life.[27,34,41-44] The majority of the cases have been acquired[34-42] and familial occurrence is only sporadic (up to 6.5%).[13-17,27,28,37,41,45,46] Autosomal dominant inheritance with variable penetrance is usual; however, autosomal recessive inheritance has also been described.[47] Monozygotic twins have been observed to develop PRP.[48] Familial PRP usually develops in childhood while acquired PRP develops in the 5th or 6th decade of life.[32-34] The development of PRP in HIV/AIDS was recognized several years after its discovery, and might show peculiarities compared to classical adult PRP; it responds to antiretroviral therapy in most instances.

CLASSIFICATION

PRP was initially classified on the basis of the age at onset, behavior, clinical appearance, and prognosis by Griffiths[34] in 1980 [Table 1]. The classical (type I) adult onset PRP shows a characteristic morphology and usually resolves in 3-4 years, whereas atypical adult-onset (type II) PRP is chronic, shows ichthyosiform and lamellar scales on the palms and soles, and alopecia of varying degrees.[2,34] The association of PRP and HIV infection has recently been identified as type VI PRP and most of the cases have been reported in young heterosexual/homosexual men.[22] It has characteristically nodulo-cystic and lichen spinulosus-like lesions, poor prognosis, and is refractory to treatment.[13-15,18-20,22,28] However, after the study of 168 Thai patients, Piamphongsant and Akaraphant[37] classified the disease into four types based on the physical findings [Table 2]. However, Griffiths'[34] classification continues to be the mainstay in practice for delineating the disease.

ETIOLOGY

The exact cause of PRP is not known-the familial type usually has an autosomal dominant mode of inheritance,[46] although recessive forms have also been recorded.[47] Genetic factors may be important; however, family history is generally not forthcoming. Epidermal hyperactivity demonstrated by a faster growth of the nails and an increase in the thymidine labeling index from a normal 3% to a high 27%, may be observed in PRP.[49-53] Finzi *et al,* observed a decreased level of serum retinol-binding protein in 11 PRP patients and their relatives,[54-58] while Frazier and Hu[59-60] and Lowenthol[61] suggested that an abnormal vitamin A metabolism and/or vitamin A deficiency may play some role in PRP etiology. However, others[62-64] did not find any decreased levels of vitamin A; thus, no correlation between vitamin A deficiency and dyskeratosis has been established.[65] Interestingly, Rothman observed that vitamin A administration has often been beneficial in follicular and nonfollicular, hyperkeratotic disease even if these diseases did not originate from vitamin A deficiency.[66]

Table 1: Pityriasis rubra pilaris-Griffith's clinical classification[34,36]

Clinical type	Lesions' distribution	Natural course	Percentage of a cases
Classical adult	Generalized	Remission in 3-4 years	55
Atypical adult	Generalized	Chronic intractable	5
Classical juvenile	Generalized	Remission in 1-2 years.	10
Circumscribed juvenile	Localized	Unpredictable	25
Atypical juvenile	Generalized	Chronic intractable	5
PRP and human immunodeficiency virus-associated type[20-22,38-40]	Face and upper trunk	Refractory	On the increase

PRP - Pityriasis rubra pilaris

Table 2: Piamphongsant and Akaraphant classification of pityriasis rubra pilaris

Types	168 patients		Clinical features
	Adult	Children	
Type I	11	21	Salmon-colored, erythematous, thick plaques on the palms and soles, extending to dorsopalmar and plantar junctions
Type II	27	59	Scaly erythematous patches on the elbows and knees
Type III	16	20	Similar patches of type II (vide supra) involving large areas of the trunk, yet not generalized
Type IV	10	4	Exfoliative erythroderma associated with diffuse follicular plugging

Furthermore, bacterial superantigens have recently been incriminated in triggering some skin diseases including juvenile PRP.[67-70] This has been corroborated by the detection of bacterial superantigens in the course of acute throat infections (*Staphylococcus aureus* and group A β *Streptococcal pyogenes*), simultaneous appearance of lesions conforming to the morphology of childhood onset/juvenile PRP, and the disappearance of lesions following administration of appropriate antibiotics. In addition, significant increases in peripheral blood mononuclear cell (PBMCs) counts against *Staphylococcal* enterotoxin B *in vitro* might suggest hyper-reactivity to some bacterial products, which may lead to childhood onset/juvenile PRP.[68]

CLINICAL FEATURES

Adult onset PRP conforms to Griffiths[34] type I classical adult and type II atypical adult classifications, the former being the most common. In contrast to childhood onset juvenile PRP,[23] adult PRP typically starts on the face and scalp and promptly spreads in the cephalocaudal direction.[2,3]

Type I classical adult onset PRP: It is characterized by follicular hyperkeratotic papules that coalesce into large, scaly, erythematous plaques, palmoplantar keratoderma, diffuse furfuraceous scaling of the scalp sometimes progressing into erythroderma.[2-4,24-28] The onset is usually acute and the eruptions begin on the head, neck and upper chest as discrete, follicular papules that often coalesce to form plaques with interfollicular erythema. The spread of the lesion is characteristically in the cephalocaudal direction. The face assumes a red-orange hue with mild to moderate ectropion. The affected skin is extremely rough to touch and feels like a file.[1] Prolonged erythema may cause resultant edema, and may precipitate a high output cardiac failure in the elderly. The palms and soles may acquire the appearance of a 'hyperkeratotic sandal', while the scalp reveals diffuse bran-like scaling. Should an erythroderma develop, a few sharply demarcated islands of unaffected skin [Figures 1A, B] are important diagnostic criteria.[2,3,71-76] Pruritus is uncommon; nail changes (if any) are marked by thickening and yellow-brown discoloration of the nail plate, subungual hyperkeratosis, and splinter hemorrhages.[2,3,77-79] Unlike psoriasis, nail dystrophy and pitting are minimal in PRP. The oral mucosa may be involved in a few patients, showing macular erythema, diffuse hyperkeratosis, and white streaks;[80] hair and teeth are normal.[2-4] Type I classical adult onset PRP runs a chronic course, three out of four cases may resolve in 1-3 years; relapses are usually uncommon.

Type II atypical adult onset PRP: It is an uncommon form of the disease that develops in middle-aged adults with atypical morphological features deviating from those described above. These patients show an admixture of follicular hyperkeratosis and lamellar scaling [Figures 2A, B] on their skin surface.[2-4,24,81] Areas of eczematous changes can sometimes confuse the clinical picture. The classical cephalocaudal progression is conspicuous by its absence; the occurrence of erythroderma is also unusual.

Type VI PRP (HIV-associated PRP): The occurrence of PRP in HIV/AIDS shows certain peculiarities[20,38-40] such as a 'filiform' pattern of keratosis on the face and upper trunk, accompanied by marked acne conglobata. This type is usually recalcitrant to conventional therapy and has a poor prognosis.[20-22,38-40] Other types are described in Table 1.

ASSOCIATED FINDINGS

Adult PRP has been found to be associated with several cutaneous and noncutaneous disorders.[82-85] the exact significance of which is a matter of speculation. The associated disorders include vitiligo, lichen planus, alopecia universalis,[12] Kaposis varicelliform eruption,[86] sero-negative arthritis,[87-90] myositis,[83] myasthenia gravis,[91] hypothyroidisim,[82] celiac sprue,[84] and other infections including HIV.[20,38-40] Infrequently, internal malignancies have been recorded in adult onset PRP.[92-95] However, prominent or increasing seborrhoeic keratoses seen in erythrodermic PRP does not necessarily imply an underlying malignancy.[85,96] An intense degree of erythema in erythrodermic PRP predisposes the individual to photosensitivity and worsening of the erythema has also been recorded with UVA and UVB.[97-99]

HISTOPATHOLOGIC FINDINGS

Adult onset PRP displays distinctive histopathological findings, which may differ according to the stage and evolution of the lesions.[2,3] The salient criteria include: (a) alternating orthokeratosis and parakeratosis in both the vertical and horizontal directions, (b) hypergranulosis, (c), irregular acanthosis apparent in the form of short and broad rete-ridges, (d) thick suprapapillary plates, and (e) a sparse to moderate lymphocytic perivascular infiltrate in the dermis[27,42,49,71,100] [Figure 3]. The hair follicles are dilated and filled with a dense, horny plug [Figure 4]. Munro's microabscesses and suprapapillary thinning are conspicuously absent. The differential diagnosis may often be difficult in erythrodermic patients.

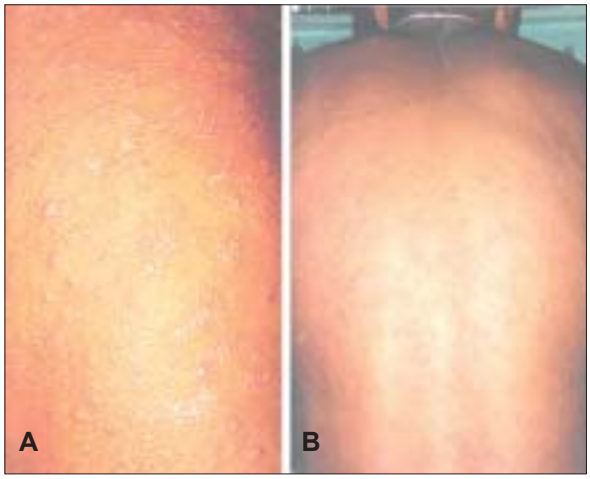
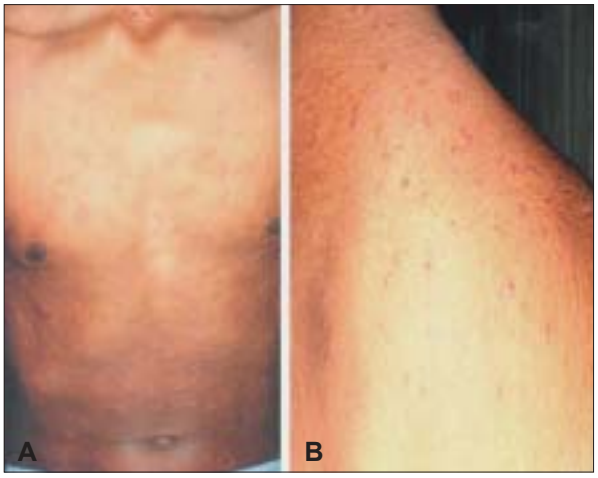


Figure 1: (A) and (B) Adult onset pityriasis rubra pilaris: Type I classical adult, multiple, discrete erythemato-scaly acuminate papules, a few coalescing to form plaques. Intervening 'islands of sparing' are conspicuous



Figures 2: (A) and (B) Adult onset pityriasis rubra pilaris: Type II atypical adult, multiple, discrete erythemato-scaly papules/plaques

Walsh *et al,*[101] found that dermatopathologist are the least (25%) accurate when scanning the sections prepared from biopsies of erythroderma of PRP origin. The dermis shows dilated capillaries with a mild to moderate infiltrate of lymphocytes and histiocytes. Acantholysis and focal acantholytic dyskeratosis have recently been recorded in adult PRP.[94,102-104] These histological parameters are unique and different from those seen in psoriasis; Magro and Crowson[43] found these features in 23 of the 32 biopsies from PRP. However, they were not found in any of the specimens of psoriasis.

Porter and Shuster[105] have been credited with the demonstration of increased epidermal replacement after they found an increase in the uptake of amino acids by the PRP lesions. Later on, several *in vivo* and *in vitro* autoradiography studies using titrated thymidine confirmed

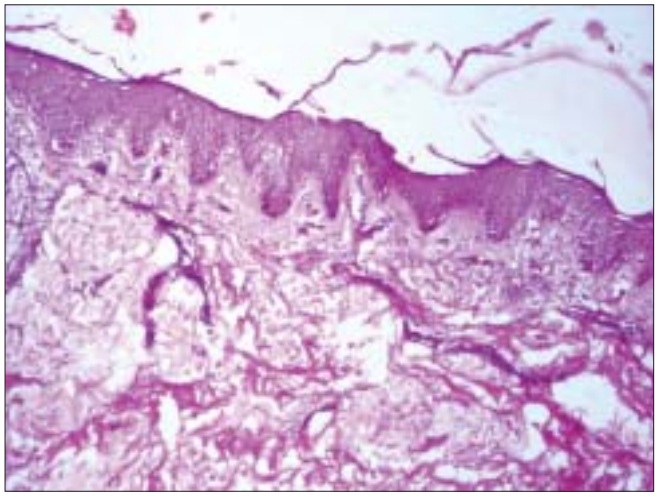


Figure 3: Adult onset pityriasis rubra pilaris: Type I classical adult, sections prepared from the biopsy of a papule (nonfollicular) depicting orthokeratosis, alternating parakeratosis, thick suprapapillary plate, short and broad rete-ridges and a mild nonspecific inflammatory infiltrate (H and E, X100)

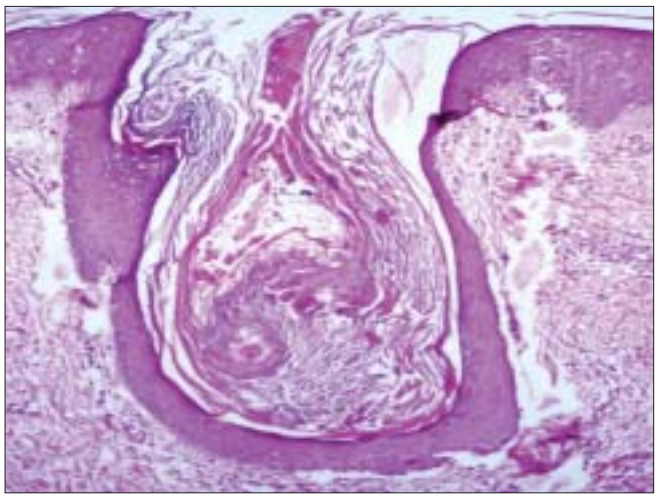


Figure 4: Adult onset pityriasis rubra pilaris: Type II atypical adult-sections prepared from the biopsy of a papule (follicular) showing in addition, keratotic plugging-an exquisite feature of the condition (H and E, X400)

an increase in the labeling index of PRP epidermal cells when compared with normal, reflecting increased cell proliferation.[42,49,50,51,53,106] Electron microscopy revealed a decrease in the number of tonofilaments, desmosomes, and enlarged intercellular spaces.[49,107] The corneocytes are fusiform and show numerous pits.[107] Evidence of parakeratosis of the stratum corneum is seen as lipid-like vacuoles, incomplete keratinization, and remnants of nuclei.[58,107]

LABORATORY FINDINGS

Hematological and laboratory test results are usually

within normal limits; the main emphasis remains on the histopathology. Plasma vitamin A and carotenoid levels are normal,[62-64] although retinal-binding protein may be low[54] or normal.[55-58] Direct immunofluorescence tests with antibodies to human IgG, IgM, IgA and complement C_3 were found to be negative in 15 adult PRP patients by Niemi

Table 3: Treatment options in adult onset pityriasis rubra pilaris

Treatments of historical importance					
Author(s)	**Years**	**Recommended drug(s)**	**Dosage**	**Response / Result**	**Number of patients**
Petter[131], Gunther[132], Gunther S, Alston W[133], Randle, Diaz-Perez, Winkelmann[134], Winkelmann, Thomas, Randle[135], Kellum[136] Murray, Gilgor, Lazarus[137] Anonymous[143]	1936, 1983, 1971	Vitamin A	1,000,000 IU/per day x/2 weeks	Good	2
Ayres, Mihan, Scribner[138]	1979	Vitamins A and E	500,000 IU + 200-400 IU	Synergism of vitamins A and E	A report
Skinner, Rosenberg, Pucevich, Kaplan [140]	1981	Cod liver oil	1-3 mL/day	Worthwhile	Case report / letter
Brunsting Sheard[141]	1941	Habibul liver oil	1-3 mL/day	Good	-
Weiner, Levin[142]	1943	Carotene	-	Good in some cases only	-
Webster and Falk[175]	1952	ACTH +Vitamin A	1,000,000 IU of Vitamin A + 10-20 units Adreno cortico trophic hormone/week	Favorable	2 patients
Irgang[178]	1968	Ascorbic acid	Oral and I/M 500 mg to 1 gm	Useful in isolated cases	-
Watt and Jilson[179]	1965	Pencillin and antitubercular drugs	Penicillin V 1 g/day plus usual anti tubercular regimen	Equivocal	6 patients
Modern treatments for adult onset PRP					
Kirby, Watson R[156], Herbst Vogelbruch, Ehnis, Kiehl, Kapp, Weiss[157]	2000 2000	Retinoid +UV light	Acetretin + UVA: 1 (0.75 mg/kg/day) acetretin + narrowband UV-B (Re-TL-01)	Favorable Favorable	Case report/ letter A case report / letter
Hofer, Mullegger, Kerl, Wolf[158] Neess, Hinrichs, Dissemond, Herrmann, Poswig, Servera-Llanras *et al*[161]	1999 2000	PUVA	Extracorporeal photo-chemotherapy	Mild to moderate good in erythroderma	Case report/ letter 1
Lamar and Gaethe[162]	1964	Methotrexate	5-30 mg/week	Equivocal good to mild	-
Duncan, Imaeda, Milstone[168]	1998				Case report/ review
Ahmed and Moy[169]	1981	Azothioprine	50-200 mg/day	Good	Review
Gendler[171]	1984				
Rosenbach and Lowe[172]	1993	Cyclosporine	< 5 mg/kg/day	Good in recalcitrant PRP	Case report/letter
Meyer and van Voorst Vader[174]	1989				case report
Lim and Tham[41]	1991	Systemic prednisolone	20-60 mg/day	Useful only in some cases	4
Brice, Spencer[55]	1985	Stanozolol	2 mg/day	Good result in some cases	Letter
Pavlidakey, Hashimoto, Savoy, Heller, Iacobelli, Barfield[180]	1985				case report
van de kerkhof and	1991	Calcipotriol	Topical ointment	Same cases	Case report/ letter
de Jong[181] Thiers [183]	1997		applied daily	may respond	
Coras, Vogt, Ulrich, Landthaler, Hohenleutner,[184]	2005	Fumaric acid	*Recommended dosages	Claimed to be useful	-
Haenssle, Bertsch, Emmert, Wolf, Zutt[185]	2004	Extracorporeal photo-chemotherapy (ECP)	2 Joule/cm^2/, monthly interval on 2 consecutive days	Good results in isolated cases	-

et al.[42] However, immunoelectrophoresis of the scales from PRP demonstrated the presence of only IgG, while psoriatic scales had IgG, IgA and C_3.[108] Takematsu *et al,*[109,110] recorded normal levels of leukotriene B_4 but low levels of anaphylotoxins in scale extracts from PRP. Other studies have been done on HLA typing,[42] direct immunofluorescence,[42] keratin monoclonal antibodies,[15] parathyroid hormone levels,[111] and a western blot analysis of the skin,[15] but these studies only have academic significance.

DIAGNOSIS

Until the disease is well-developed, it may be difficult to diagnose with full confidence. However, repeated observations and a few biopsies may confirm the diagnosis.[2-4,24,112-116] Atypical (type II) PRP may be more difficult to diagnose than classical adult onset (type I) PRP. Follicular hyperkeratosis on the back of the fingers, orange-colored eruptions with intervening areas of normal skin 'islands of sparing' and/or palmoplantar keratoderma are features of classical adult onset PRP; they are ill defined in atypical adult PRP.[2-4,117] The differential diagnosis of adult onset PRP usually includes psoriasis.[2,4,118] The absence of Auspitz and candle grease signs is an instant clinical diagnostic clue.[2,3] Erythrodermic PRP can be confused with other forms of erythroderma[119] and skin biopsy of such patients can confirm PRP only on an exclusion basis.[101] Arthropathic PRP is unusual to record.[120] Resolving PRP may mimic seborrhoeic dermatitis[2] or erythema gyratum repens.[17,121] PRP may be a cutaneous marker of internal malignancy,[92,93,95,122] leukemia,[93] metastatic carcinoma,[92] or Sezary syndrome[123] in adults, which may follow after a variable length of time. Interestingly, cutaneous T-cell lymphoma and Sezary's syndrome also form a differential diagnosis of erythrodermic PRP.[124,125] Rarely, dermatomyositis may develop skin eruptions akin to the adult onset PRP.[126-130] Heteroduplex analysis of T-cell receptor gamma gene arrangements may be a newer adjuvant diagnostic tool in skin biopsies from erythrodermas.[128]

TREATMENT OPTIONS

The diagnosis and treatment of PRP have always been a source of great interest. There is no acclaimed treatment for PRP at present. Thus, affected individuals often visit and change many treating dermatologists to alleviate their signs and symptoms. More often than not, it is an exercise in futility as the treating physician/dermatologist too is in dilemma. Several treatment[131-191] options have been *in vogue* and are tabulated below [Table 3].

Narrowband UV-B with oral retinoids has been useful in some cases.[182,187] Topical calcipotriol[138] and tacalcitol have also given promising results in some patients. HIV-associated PRP is more recalcitrant but antiretroviral drug therapy has caused alleviation of the symptoms and may even cause complete regression in such patients.[188] Methotrexate has been found to be moderately effective. In an attempt to explore an ideal therapy, newer treatment options like biologicals (infliximab), calineurin inhibitors (pimecrolimus) etc. are being tried in PRP.[192,193] The use of emollients to symptomatically improve the condition may also be useful. It is imperative to record at this point in time, that several treatment options that have been used so far may not be satisfactory as no organized drug trials are available. Nevertheless, isotretinoin, a retinoid, seems to be a plausible option.[149,154,189-191]

The historical and epidemiological perspectives of adult onset PRP as well as its etiology have been described. Microscopic pathology and its variations have been clearly defined, emphasizing its role in supplementing clinical diagnosis and treatment has been facilitated by the inclusion of a table for decision-making.

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