

ンケート調査を行い、容姿による不幸福感、自信喪失、いじめなどが手術に踏み切った主要因であること、容姿が最も QOL を下げていること、全例が追加手術を検討しており、他の患者にも手術を勧めたいと回答していることを報告している。

整容面の改善度を客観的に評価することは困難であるが、手術の主目的は患者の精神的な苦痛を取り除くことであり、その点からは美容外科手術は有用な治療の選択肢の一つと考えられる。なお、術後に再燃する例も報告されており、疾患活動性が十分落ち着いてから手術に踏み切るのが適切と考えられる。

CQ22 脳病変に対して有用な治療はあるか？

推奨文： 脳病変によって生じる軽症のてんかん発作には抗てんかん薬を推奨する。

活動性のある脳病変に全般性強直間代発作あるいは治療抵抗性のてんかん発作など中等症以上のてんかんを伴う場合は、ステロイド全身療法と免疫抑制薬の併用を選択肢の一つとして提案する。

推奨度： 抗てんかん薬：1D、ステロイド全身療法と免疫抑制薬の併用：2D

解説：

限局性強皮症において最も高頻度に見られる脳神経症状はてんかんであり、各種抗てんかん薬がてんかん発作の治療に用いられている¹⁴¹⁻¹⁵¹。カルバマゼピン、オキシカルバゼピン、フェノバルビタール、バルプロ酸ナトリウム、トピラマート、クロバザム、プレガバリン、ニトラゼパム、ビガバトリン、スルチアム、ラモトリギンなどが使用されており、軽症例では抗てんかん薬は78%の症例において、てんかん発作のコントロールに有用であったと報告されている¹⁵²。

全般性強直間代発作あるいは治療抵抗性のてんかん発作には、免疫抑制療法が必要である。最も高頻度で使用されているのは副腎皮質ステロイドで過去の報告例では80%で使用されており、90%の有効率と報告されている^{127, 142, 143, 145-147, 153, 154}。しかし、副腎皮質ステロイド単独で治療されているのは1例のみであり¹¹¹、メソトレキサートを併用した症例が4例^{142, 143, 146, 154}、アザチオプリンを併用した症例が3例^{125, 129, 131}、シクロホスファミドを併用した症例が2例¹⁴⁷、ミコフェノール酸モフェチル¹⁴²、インターフェロン- γ ¹⁴⁵、免疫グロブリン大量静注療法¹⁵³を併用した症例が1例ずつ報告されている。免疫グロブリン大量静注療法以外では症状の改善がみられたと報告されている^{153, 155}。

持続性部分てんかんに機能的な脳半球切除術を施行した1例¹⁵⁶、治療抵抗性のてんかん発作に対して部分皮質切除を施行した2例が報告されており^{153, 157}、いずれも症状の改善がみられたとされている。その他、進行性多巣性白質脳症、脳卒中、末梢神経障害が副腎皮質ステロイドで良好にコントロールできた症例が報告されている¹⁵⁸⁻¹⁶⁰。また、不全片麻痺については、1例はステロイドパルスが有効、1例はステロイドパルスと免疫グロブリン大量静注療法が無効であったと報告されている^{153, 161}。再発性頭痛や脳神経病変に対して副腎皮質ステロイド、メソトレキサート、ミコフェノール酸モフェチルが有効であった症例が報告されている^{142, 143, 162, 163}。また、抗けいれん薬や抗うつ薬が症状の軽減に有効であったという報告もある^{144, 164, 165}。視神経乳頭炎に対して、副腎皮質ステロイドやアザチオプリンが無効であった症例の報告がある¹⁴⁵。脳血管炎が1例で報告されているが、MMFで改善している¹⁶²。

以上より、脳病変によって生じる軽症のてんかん発作には抗てんかん薬が有用であり、全般性強直間代発作あるいは治療抵抗性のてんかん発作など中等症以上のてんかんには、ステロイド全身療法と免疫抑制薬の併用が有用であると考えられる。なお、抗てんかん薬については、エビデンスレベルは低いだが

イドライン作成委員会のコンセンサスのもと、推奨度を 1D とした。

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Ⅲ. 好酸球性筋膜炎の診断基準・重症度分類・診療ガイドライン

1. 診断基準

好酸球性筋膜炎の診断基準

大項目 四肢の対称性の板状硬化

但し、レイノー現象を欠き、全身性強皮症を除外しうる

小項目 1 筋膜を含めた皮膚生検組織像で、筋膜の肥厚を伴う皮下結合織の線維化と、好酸球、単核球の細胞浸潤

小項目 2 MRI 等の画像検査で筋膜の肥厚

大項目及び小項目 1 ないし 大項目及び小項目 2 で診断確定

Ⅲ. 好酸球性筋膜炎の診断基準・重症度分類・診療ガイドライン

2. 重症度分類

好酸球性筋膜炎の重症度分類

- | | |
|------------------------|-----|
| ・ 関節拘縮を伴うもの（上肢） | 1 点 |
| ・ 関節拘縮を伴うもの（下肢） | 1 点 |
| ・ 運動制限を伴うもの（上肢） | 1 点 |
| ・ 運動制限を伴うもの（下肢） | 1 点 |
| ・ 皮疹が拡大増悪（症状が進行）しているもの | 1 点 |

点数を合計して2点以上は重症

3. 診療ガイドライン

好酸球性筋膜炎の診療アルゴリズム

好酸球性筋膜炎の診療アルゴリズム

